UK Forum on
Haemoglobin
Disorders

WM QRS West Midlands
Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia
at
Mayday Healthcare NHS Trust

Quality Review Visit Report
Visit date: Wednesday May 19th 2010

Report finalised: July 29th 2010

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INTRODUCTION

This report presents the findings of a peer review visit to services for children with sickle cell disease or thalassaemia at Mayday Healthcare NHS Trust which took place on May 19th 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. Although Mayday Healthcare NHS Trust was not on the original list of 'centres' to be visited, on discussion it became apparent that it was a providing a large and significant local service and warranted a separate visit. The visit to Mayday was a half-day visit, with a smaller review team than the 'centre' visits, but reviewers felt that it had been possible to discuss and evaluate the important aspects of the service during the visit.

ACKNOWLEDGEMENTS

We would like to thank the staff of Mayday NHS 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 MAYDAY HEALTHCARE NHS TRUST

Service (as at May 2010)	Patient numbers sickle cell disease	Patient numbers thalassaemia	Patients on long term red cell transfusions
Mayday Hospital	Total 214 < 19yrs [138 SS, 51 SC]	<5	8 [<5 transfused here]

Mayday Healthcare NHS Trust [Mayday] is sited in Croydon, which is a densely populated borough. A high proportion of resident families are black or of minority ethnic group. The hospital provides a comprehensive local service for children with sickle cell disease, of whom a total of 214 are known to the service. 165 receive all their regular care at Mayday. Fewer than five children with thalassaemia syndromes attend the hospital, and a minority of these have regular transfusions at this hospital. Of eight children with sickle cell disease [<five post – stroke] who receive regular red cell transfusions, a minority have chosen to attend Mayday for these. Fewer than five children are treated with

hydroxycarbamide. An average of 12 affected newborn were joining the service each year. There are close working links with King's College Hospital [KCH].

There are a total of eight paediatricians; one is the named lead for haemoglobinopathies. For two weeks in six he manages the neonatal unit, for the remaining four weeks he spends approximately three days per week on the red cell services. Medical cover for in-patients is by paediatric consultant colleagues. The adult haematologist with an interest in red cell disorders does not have regular input to the care of these children in the out-patient or in-patient setting. There is a strong community nursing team, with clinical nurse specialist having particular responsibility for the children using this service. She sees children when they are in-patients and provides very useful continuity and specialist guidance. Advice is readily sought from the paediatric haematologists at KCH, who now visit monthly to see children in clinic with the lead paediatrician. The small number of children receiving regular transfusions / chelation are jointly managed. Annual review of other children takes place at these clinics and trans-cranial Doppler screening for stroke risk assessment is also offered.

A regular sickle cell clinic was started in 2005, and runs weekly except for the two weeks in six when the lead paediatrician is managing the neonatal unit. The community nurse attends these clinics. The outreach clinic, when one or other of the two paediatric haematologists from KCH come to Mayday, continues every month whether the lead paediatrician is there or not. Visiting tertiary specialists also hold endocrine and respiratory clinics to which these children can be referred and seen on site. There is a visiting children's cardiac technician. Children are usually referred to Evelina Children's Hospital for speciality input for renal, cardiac, or urological problems. Support is available from the play therapist, dietician and social worker as needed. Psychology services can be accessed through the local CAMHS or by referral to KCH. Transition clinics, run by the lead paediatrician and nurse specialists, take place as needed when there are children coming up to the age of transfer, which can be up to 18 years.

Children needing urgent assessment and care present to the A&E Department, where they are 'fast triaged' through to the children's area. Children with sickle cell have a blue 'dot' added to their A&E card so that staff know they will need prompt attention. A paediatric registrar is in A&E from 11 am – 8 pm. In the morning a more junior paediatric trainee is present and overnight cover is from the on-call paediatric registrar. Initial analgesia, depending on pain score, is usually oramorph, with move to parenteral morphine if pain does not settle. There are approximately 55 admissions for sickle cell complications per year.

Admission is to a ward area where staff are familiar with these conditions. Clinical guidelines are available on the ward together with some relevant research articles for nurses' interest and education. Older children tend to be cohorted in one of the four-bedded bays. If children are acutely ill, or need exchange blood transfusion, they are transferred to KCH. Children needing PICU are retrieved and taken to the nearest available bed.

Day care is offered flexibly, on a separate day unit, according to school timetables, but out of hours transfusion is not available. Cannulation is either by the nurse phlebotomist in out-patients, or by a paediatric doctor.

Community services are very strong and active, running popular support groups and age-cohorted sessions for children and teenagers. There is much health promotion and fund raising work. The lead community CNS for this service 'in-reaches' to a large extent and can cover some of the roles of an acute hospital based CNS which the service currently lacks. Cover for her is from two colleagues, who have responsibility for screening services, and adults respectively.

Communication with the centres and other hospitals in South London is felt to be good, with three to four times yearly meetings of the 'SKILS' group, which addresses network issues, and is developing shared guidelines. The monthly contact with one of the visiting paediatric haematologists from KCH additionally gives an opportunity for case discussion.

REVIEW VISIT FINDINGS

ACHIEVEMENTS

1 This is an excellent local service, which ensures that almost all key clinical standards are met by the efforts and energies of the small local team supported by ready access to, and regular outreach by, members of the specialist team at KCH. The establishment of a regular on-site TCD screening, and systematic annual review of patients by the visiting specialists, is very good.

2 The activities of the small community nursing team are truly outstanding, and the service as a whole and the community nurse support especially, is highly valued by users.

3 The facilities from which these services are offered are of a high standard, and the efforts of staff to make the best use of them are clear.

4 TCD coverage is very high.

IMMEDIATE RISKS

No immediate risks were identified.

CONCERNS

1 User families who met the review team reported that the fast track system in A&E does not work well for them. Sometimes they have to wait in a queue in the reception area to be 'booked in' and reception staff can be unhelpful or even aggressive.

FURTHER CONSIDERATION

1 Medical and specialist nursing staff numbers for this large a service are low. This would have been of concern only but for the efforts and commitment of the staff who are in post, the willingness of the other staff to cover and the specialist support offered by the King's College Hospital outreaching for a joint monthly clinic. The service is clearly growing, and is likely to continue to do so. It is hard to see how all the children can be seen as often as needed in a weekly clinic, run by a single doctor, four weeks out of six.

2 Given the concerns of families about waits to access care in A&E, an audit of timelines for children attending to time assessed, time to first analgesia, and time to adequate analgesia, may be useful.

3 Out of hours transfusion facilities would be useful. One teenager currently receiving his transfusions here is considering moving elsewhere as this is not available. The ward staff do make efforts to adapt to the convenience of the patient as far as they are able and this is noted and appreciated by the families.

4 Given the small number of clinical staff, help with data handling will be needed if the service is to keep adequate data to inform practice audits, and to enter large numbers of patients onto the National Haemoglobinopathy Registry.

5 As only a very small number of children with thalassaemia attend Mayday, it may be helpful to offer these families transfer to another hospital in the area with more patients for reasons of peer and family support. Families may not wish to make this transfer, but they should be made aware of other larger units that they might access for clinical or support activities.

6 Play therapy is not easy to access with one therapist covering all clinical areas. Psychology can be accessed but is not readily available and so children are currently only referred if there are significant problems. The level of support recommended for children and families with these conditions is not currently possible.

GOOD PRACTICE

1 Many of the clinical guidelines are noteworthy, in particular the nursing guidelines for care, the A&E clinical assessment sheet, and the pain management flowchart which is clear and easy to follow.

2 The quality of much of the information for patients and families is very high. A full range of material is available for those affected by thalassaemia, as well as for those with sickle cell, which is commendable considering the much smaller numbers of patients.

3 The personalised 'school packs' to facilitate communication between health services and schools are excellent.

4 The offer of home visits by the CNS to discuss any difficult or sensitive issues with children, especially around transition age, is commendable.

5 The flexibility for children to remain under paediatric services up to age 19 is good.

6 User feedback about the quality of care and support they receive overall is very positive.

7 It is helpful, and unusual, to have a children's social worker on site contributing to the service.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Lee McPhail	Assistant Director of Operations	North Middlesex University Hospital
Elaine Miller	User Representative	UK Thalassaemia Society
Dr Anne Yardumian	Clinical Lead for Peer Review Programme	

APPENDIX 2: COMPLIANCE WITH QUALITY REQUIREMENTS

Local 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	*These materials are excellent and comprehensive. It is not clearly documented who the 'key contact' is, but in discussion the community CNS confirmed that her team issue contact 'cards' to all families, indicating that they are the point of first contact within working hours, in emergency or out of hours, they are recommended to go straight to A&E.*

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).	Y	*This is very good material.*
5	The locally agreed patient-held record (QR4) should be in regular use within the LHT / SHT.	N	*No patient held record in systematic use.*
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 clinical areas are spacious and light and child and family-friendly.*
STAFFING and SUPPORT SERVICES			
7	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	
8	There should be agreed arrangements for cover for absences of the lead paediatrician.	Y	
11	The LHT should have a lead nurse who has responsibility, with the lead consultant (QR7), for guidelines, protocols, training, audit relating to haemoglobinopathies, liaison with community services and liaison with the SHT. The lead nurse should have specific training in the care of patients with haemoglobinopathies.	Y	
17	The following support services should be available: a Interpreters b Social work c Play specialist d Hospital teacher (in-patient care only) e Child psychologist f Dietician	Y	*But full psychology services are not sufficiently readily available and play support is sparse.*
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	*But working between the community and acute services is very well integrated in practice and the community team offers more input than in many other areas.*
19	A nurse with competency in cannulation, starting and supervising a transfusion should be available at all times at which children are attending for transfusion.	N	
20	All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training.	Y	

Ref	Quality Requirement	Met?	Comment
CLINICAL and REFERRAL GUIDELINES			
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 are excellent.*

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	
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	N	*} There are some general hospital transfusion guidelines but these*
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.	N	*} were not seen so it is not clear if*

Ref	Quality Requirement	Met?	Comment
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.	N	*} they cover the content of these requirements.*
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.	N	*No guidelines seen.*
33	Clinical guidelines should be in use covering: a Indications for exchange transfusion b Arrangements for carrying out an exchange transfusion.	N	*But this is not done on this site.*
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)	Y	
36	A protocol should be in use covering the initial clinic visit for patients previously treated outside the UK, including: a Full medical history and examination b Investigations c Referral to other specialist services (QR15) d All aspects of QR28	N	

Ref	Quality Requirement	Met?	Comment
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	
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	
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	
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	
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	*Much in evidence.*
46	Staff from the LHT should participate in the training and awareness programme run by the SHT to which patients are usually referred.	Y	
48	A representative of the LHT should attend each review meeting with the SHT to which patients are usually referred (QR47).	Y	

Ref	Quality Requirement	Met?	Comment
DATA COLLECTION and AUDIT			
50	The LHT / SHT should have audited compliance with key standards including: **Patients with sickle cell disease:** a Proportion of patients taking regular penicillin b Proportion of patients fully immunised against pneumococcus c Proportion of patients (HbSS and HbSβ^0) who have had Transcranial Doppler ultrasonography undertaken within the last year d Proportion of patients who have had their annual multi-disciplinary review within the last year e Effectiveness of action to contact families who have not attended for follow up appointments. f Review of the care of any patients who have died. **Patients with thalassaemia:** a Proportion of patients on chelation therapy b Proportion of patients who have had their annual multi-disciplinary review within the last year c Adequacy of recording of: a. Pre-transfusion Hb levels b. Regular monitoring of iron level c. Complications of iron overload d. Height / weight progression e. Spleen size f. 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.	Y	*Audit data presented referred to a relatively small number of children [47] with sickle cell disease, but it was explained in discussion that these were all children under 3 years of age.*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	Y	*Although only 10 patients to date.*

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.		*There was no discussion at this visit about commissioning arrangements.*

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.		*As for 52.*

Additional Requirement – Screening Services

Ref	Quality requirement	Met?	Comment
P3	Timely communication of positive screening results (sickle cell disorder) – including a review of parental results	N	*But data provided suggests that where the screening result was received in time, this standard 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)	N	*Approximately 57% babies seen by 3 months, and 11 children not seen until > 20 weeks.*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	Y	
S1i	Failsafe to ensure ongoing care	Y	*The community nurse described detailed steps taken, although in practice this is rare for new families.*
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.