UK Forum on
Haemoglobin
Disorders

WM QRS West Midlands
Quality Review Service

Services for Children with Sickle Cell Disease or Thalassaemia
at
Queen Elizabeth Hospital, Woolwich

Quality Review Visit Report
Visit date: Wednesday May 19th 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 Queen Elizabeth Hospital 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 Queen Elizabeth Hospital 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 Queen Elizabeth Hospital 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 Queen Elizabeth Hospital for their hard work in preparing for the review and for their kindness and helpfulness during the course of the visit. 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 QUEEN ELIZABETH HOSPITAL

Service (as at May 2010)	Patient numbers sickle cell disease	Patient numbers thalassaemia	Patients on long term red cell transfusions
QE Woolwich	204 [141 SS, 43 SC, 8 other]		<5

Queen Elizabeth Hospital [QEH] is one of three hospitals which together make up the South London Healthcare NHS Trust. The others are Queen Mary's Sidcup Hospital and Bromley Hospital. The area has a high and rising black and minority ethnic population, and a high birth rate. There were 20 babies with sickle cell disease identified through the newborn screening programme and referred to QEH in 2009, and 14 in the first five months of 2010. Queen Mary's Sidcup looks after approximately 100 children with sickle cell disease and a small number with thalassaemia. Bromley Hospital looks after about 14 with sickle cell disease.

The lead paediatrician for this service has been in post for three years. She is also lead for epilepsy and has a number of other specific departmental responsibilities. She, together with specialist community nurses, currently constitutes the team managing these children across all clinical settings. There is no specialist nurse for the acute hospital service. There is some support from the specialist paediatric haemoglobinopathy team at King's College Hospital [KCH] who for the last two years has undertaken outreach joint clinics at QEH and are available for discussion or referral of difficult individual cases.

There is currently a major shortfall in paediatric medical trainees, four of nine specialist training posts were unfilled and one of two staff grade posts was unfilled. A series of shorter or longer-term locums were filling the posts. There is also a shortfall in the community nursing team. The Greenwich sickle cell and thalassaemia centre had four specialist nurses, employed by Greenwich PCT, working across Greenwich and Bexley and, until recently, Bromley. The team's duties are:

- to undertake antenatal counselling and couple counselling for those at risk of having an affected baby
- to receive and communicate the results of the newborn screening programme, visiting families of affected newborn at home to give the diagnosis and early support and education
- to refer to the hospital clinic
- to work in the weekly clinics with the lead paediatrician and haematologist, to input to the care of patients in hospital
- to offer school liaison and other support to children of all ages, and provide a range of services for the adult patients.

In principle the same nurse works in one borough through to the adult service, facilitating transition. The nurses also run an all-age support group, which is held in an evening once per month and is well attended. There are currently two vacant posts, and no plans to re-appoint to one of these. The community nurse who leads for children and adults in Greenwich has had to pick up antenatal and newborn counselling. This is a particular problem as the antenatal clinic and weekly sickle cell clinic are held at the same time.

The lead paediatrician runs a weekly clinic for children with sickle cell disease, usually together with the community nurse with responsibility for children in Greenwich. The DNA rate has been very high but efforts made by the community nurse to remind families of their appointments have brought this

to near zero. There is no phlebotomy service in the clinic. Younger children come to a separate nurse phlebotomy clinic, where there is play input. Children over 3 years old use the general phlebotomy area. Social services and dietary support are available by referral. Psychology is provided by CAMHS but in practice is only offered to children who have quite major problems. There is a 3 monthly transition clinic held with the haematologist leading the adult red cell service. On one morning per month, a paediatric haematologist from KCH comes to see children jointly with the QEH lead. Annual review on some of the children is undertaken and experienced KCH staff bring a portable Doppler machine for trans-cranial Doppler screening for stroke risk assessment.

There is a semi-formal regional network, which holds three to four monthly education and management meetings, the 'SKILS' group. The QEH paediatrician refers for tertiary care both to Evelina Children's Hospital [ECH] and KCH for different clinical problems. Children are referred to ECH for cardiac, renal, neurological, and urological complications and to KCH for neurovascular work, laparoscopic abdominal surgery and orthopaedics. There is no respiratory consultant in the network managing sickle lung problems.

Children needing urgent care are asked to come to paediatric A&E, which is open from 8 am to 2 a.m. Between 2 and 8 am the adult A&E is used. Children have to use the main reception area. Triage is reported to be usually within 15 minutes. The medical staff in paediatric A&E is mostly not paediatric-trained, and the posts are quite frequently covered by locum doctors. The Trust intend that there should be paediatric trained nurses on every shift. Complex patients carry a 'passport' indicating that they should be referred for assessment to the paediatric team. Analgesia is usually with oral analgesia up to morphine as necessary, and parenteral morphine only if oral is not proving effective. No individualised care plans are held. Nursing staff decide with parents what to use, depending on what has already been given. The lead paediatrician frequently undertakes teaching for A&E staff about acute sickle cell presentations. From September 2009 the A&E Department at Queen Mary's Sidcup is closed from 8 pm to 8 am. One to two children per week are admitted for sickle cell related problems.

Admission is to a pleasant in-patient facility. This currently has 16 beds which the lead clinician has been informed is soon to expand to 34. Staff are familiar with nursing children with sickle cell disease, and some clinical guidelines are available on the intranet. There are 'high dependency' beds close to the nursing station in which some children, for example those needing morphine infusions, are managed. There is input from the pain team when morphine nurse or patient controlled analgesia pumps are in use. Children requiring red cell exchange are discussed with and referred to the specialist team, usually at KCH.

Day care, including booked transfusions, takes place in a large ambulatory care unit, seeing approximately 300 children per month. It is used also for children coming from A&E who may need a slightly longer stay. It is unusual for elective transfusions to be cancelled because of bed availability. Some of the unit nurses can cannulate, otherwise a paediatric doctor is called. Transfusions are prescribed in advance. At present only a small number of children receive their regular transfusions here.

Review Visit Findings

Achievements

1 This is a large and growing clinical practice, delivered by a small clinical team working very hard to develop the service. Some aspects of the service are working well, for example, the on site outreach specialist clinics and TCD service. There is close team working between the acute and community service. The energy and enthusiasm of the lead paediatrician is evident and she has many ideas and plans as to how to improve the service. The contribution of the community nurse team is very valuable in the acute setting, in the absence of an acute hospital-based CNS.

2 The community nurses have been delivering a wide range of services across a wide geographical area. They are obviously highly committed to the service although, if their numbers are reduced, their ability to continue working in this way will be affected [see 'concerns'].

3 The reduction of clinic DNA rates from up to 50% to near zero is a major achievement.

4 The facilities from which the services are delivered are of a high standard.

Immediate Risks

No immediate risks were identified.

Concerns

1 There are no clinical guidelines available in the clinical areas for some of the most frequent and serious acute paediatric sickle cell presentations. Given the number of temporary staff in

A&E and in the paediatric medical teams, some clear written guidance should be put in place as soon as possible.

2 This service is substantially under-resourced, in medical and specialist nursing provision. The lead paediatrician has many other demands on her time. With the changes in emergency care facilities at Queen Mary Sidcup, there are likely to be more families coming to QEH for their care. There is also a striking increase in numbers being identified through the newborn screening programme. The visiting team was not made aware of any plans to increase the staffing to meet the growing demand for care.

3 The community specialist nursing team covers a wide range of clinical responsibilities, both in the hospital and in the community across a large geographical area, and apparently staff numbers are to be reduced. If this happens, it will have a major adverse impact on this service. The visiting team hoped that further consideration will be given to the proposed changes, given the continued growth in service need for this patient group.

4 As well as clinical guidelines for management of common presentations, there should be clear guidance for A&E and junior paediatric medical staff as to when to escalate for advice from the on call QEH consultant, who should have guidelines on when to ask for advice or refer the child on to KCH for specialist management.

5 There was no evidence of Trust or PCT awareness of the difficulties facing this service or the expected growth. No plans for addressing the issues identified in this report were evident.

FURTHER CONSIDERATION

1 An audit of time of arrival for children presenting with acute pain to A&E, through triage to being assessed, receiving first analgesia, and time to effective pain control, should be considered given some of the concerns about staffing in A&E.

2 There is the potential for further joint work between three hospitals in the South London Healthcare NHS Trust about how best to manage patient flows and organise staffing arrangements for this service.

3 Limited psychology input is available as only children with significant problems receive this service. Children are not seen by a psychologist in the systematic way recommended, including neurocognitive assessments.

4 Some assistance with data handling will be required if this team are to collect data required for clinical audit, and enter children onto the National Haemoglobinopathy Registry. It is commendable that the lead paediatrician has managed to enter 50 children so far.

5 There are some issues of document control, with some guidelines being undated, without author or review date, and others apparently still in use while well past their intended review date. (For example, an ICP for children receiving transfusion was dated 2003, with planned review date 2004. The visiting team were subsequently informed that this is being updated and a final draft is being circulated before final approval, expected before the end of 2010).

6 The lead paediatrician would value sharing of guidelines across the network, which should be possible and would allow full guidelines to be in place more quickly, after adaptation for local use.

GOOD PRACTICE

1 Babies born at QEH who are known to be at high risk of having sickle cell disease are tested at birth and these results are used to start treatment early, and to inform families. This allows for early communication to families, who may be anxiously awaiting a result, and starts the process of referring to clinic before the results of the newborn heel-prick screen are available.

2 The lead paediatrician has an active teaching programme about haemoglobinopathies for various key clinical staff groups in the hospital.

3 There is a 'buddying' scheme for families of affected newborn, so that more experienced parents can support and guide new parents; this is in addition to an active support group led by two of the community nurse specialists.

4 Some of the guidelines are particularly helpful, including one for the management of pain in children over one year old in A&E.

5 The presence of a play specialist in outpatient clinics and in the under three year old phlebotomy clinic is commended.

APPENDIX 1: MEMBERSHIP OF THE REVIEW TEAM

Lee McPhail	Assistant Director of Operations	North Middlesex University Hospital NHS Trust
Elaine Miller	Co-ordinator	UK Thalassaemia Society
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	*Material for sickle cell disease and thalassaemia were presented.*

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	*No specific material for older children or about transition to adult services was seen, although transition clinics take place approximately quarterly, run by the lead paediatrician and the adult haematologist who manages the red cell services, usually with one of the nurse counsellors also. Patients are given a copy of their own record at these appointments.*
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	*All fine*
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.	N	*Specific guidance to covering general colleagues about when to seek advice / refer out should be in place.*
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.	N	*In reach activity by one of the community specialist nurses helps fill this gap, although that team may be reduced in numbers which would allow her to undertake less of this hospital based work.*
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	*Full psychology services to the recommended level are not readily available.*

Ref	Quality Requirement	Met?	Comment
18	A service agreement for support from community services should be in place. This service agreement should cover, at least: a Guidelines for involvement of community paediatric services in the care of patients with haemoglobinopathies b Role of community services c Exchange of information between hospital and community services and vice versa d Arrangements for liaison with schools.	Y	*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	*An old ICP [review date 2004] was included in evidence, it is apparently being updated but does not cover the content of this QR.*
20	All staff involved in the care of children and young people with haemoglobinopathies should undertake regular child protection training.	Y	*Training records for key staff were forwarded after the visit.*
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	*These are in place, though undated, and were sparse on detail.*
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	N	*Some are in place and on intranet but others – including chest syndrome, stroke, splenic sequestration, abdominal pain - were not available.*

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	*These were seen, although they lacked detail.*
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	
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	
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	*This is covered in the ICP for children receiving transfusion but dated 2003, review date 2004.*

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	*As for QR 29.*
31	Clinical guidelines for chelation therapy and monitoring iron load should be in use including: a Indications for starting chelation b Choice of regime c Dosage and dosage adjustment d Clinical assessment of tissue damage e Monitoring of serum ferritin f Use of non-invasive estimation of organ-specific iron loading heart and liver by T2*/R2 g Management of side effects of chelators.	N	*Most decisions about chelation are taken in conjunction with the specialist team.*
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	
33	Clinical guidelines should be in use covering: a Indications for exchange transfusion b Arrangements for carrying out an exchange transfusion.	N	*Exchange transfusions are 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	*Full guidelines were not in place in any location.*
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	
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	*Although self assessed as 'no' there is evidence of participation in network meetings and the specialist team outreaching on a monthly basis gives additional opportunity for discussion.*
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:** 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.	N	*No audit data relating to these standards was presented.*
51	Data should be systematically entered on all patients, following patient / parental consent, onto the National Haemoglobinopathy Registry.	Y	*Entry to NHR is well underway.*

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	*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.	N	*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	Y	*Early testing of babies at risk allows diagnosis and family visit to inform ahead of the newborn screening programme result.*
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	*100% of babies identified by newborn screening in the area were seen by 3/12 [11-baby audit presented, May 2009 - Jan 2010]*
P5	Timely confirmation of diagnosis for infants with a positive screening result**	Y	
S1i	Failsafe to ensure ongoing care	Y	
S1ii	Up-to-date registers maintained of babies (cases) for which units are responsible	Y	

** Specified conditions: Hb-SS, Hb-SC, $HbSD^{Punjab}$, Hb-SβThalassaemia (β^{+}, β^{0}, δβ, Lepore), $Hb\text{-}SO^{Arab}$, Hb-S/HPFH

NOTE. Different QR's are not comparable in terms of their importance or likely impact on the quality or outcomes of the service, and a figure summarising the number of QR's met cannot be used to assess the overall standard of the services provided or to compare this Trust's services with others.