Health Services for People with Haemoglobin Disorders

Alder Hey Children's NHS Foundation Trust

Visit Date: 21st June 2019

Report Date: October 2019

8831

Contents

Introduction

This report presents the findings of the peer review of health services for people with haemoglobin disorders in Alder Hey Children's NHS Foundation Trust that took place on 21st June 2019. The purpose of the visit was to review compliance with the Quality Standards for Health Services for People with Haemoglobin Disorders V4, 2018 which were developed by the UK Forum on Haemoglobin Disorders working with the Quality Review Service (QRS). The peer review visit was organised by QRS on behalf of the UK Forum on Haemoglobin Disorders. The Quality Standards refer to the following types of specialised service for people with haemoglobin disorders:

- Specialist Haemoglobinopathy Team (SHT)
- Local Haemoglobinopathy Teams (LHT) or linked providers

Haemoglobin Disorder Network and Commissioning Quality Standards were not reviewed as part of the 2019-20 review programme, although reviewers did enquire about the local network and commissioning arrangements during the course of the review visit.

The aim of the standards and the review programme is to help providers and commissioners of services to improve clinical outcomes and service users' and carers' experiences by improving the quality of services. The report also gives external assurance of the care which can be used as part of organisations' Quality Accounts. For commissioners, the report gives assurance of the quality of services commissioned and identifies areas where developments may be needed.

The report reflects the situation at the time of the visit. The text of this report identifies the main issues raised during the course of the visit. Any immediate risk identified includes the Trust's proposed actions to mitigate the risk and QRS's response to those proposals. Appendix 1 lists the visiting team that reviewed the services at Alder Hey Children's NHS Foundation Trust. Appendix 2 contains the details of compliance with each of the standards and the percentage of standards met.

This report describes services provided or commissioned by the following organisations:

- Alder Hey Children's NHS Foundation Trust
- NHS England Specialised Commissioning – Haemoglobinopathies

Most of the issues identified by quality reviews can be resolved by providers' and commissioners' own governance arrangements. Many can be tackled by the use of appropriate service improvement approaches; some require commissioner input. Individual organisations are responsible for taking action and monitoring this through their usual governance mechanisms. The lead commissioner for the service concerned is responsible for ensuring action plans are in place and monitoring their implementation liaising, as appropriate, with other commissioners, including commissioners of primary care. The lead commissioner in relation to this report is the NHS England & NHS Improvement Lead Commissioner Haemoglobinopathies.

About the Quality Review Service

QRS is a collaborative venture between NHS organisations to help improve the quality of health services by developing evidence-based Quality Standards, carrying out developmental and supportive quality reviews (often through peer review visits), producing comparative information on the quality of services and providing development and learning for all involved.

Expected outcomes are better quality, safety and clinical outcomes, better patient and carer experience, organisations with better information about the quality of clinical services, and organisations with more confidence and competence in reviewing the quality of clinical services. More detail about the work of QRS is available at www.qualityreviewservicewm.nhs.uk

Acknowledgments

We would like to thank the staff of Alder Hey Children's NHS Foundation Trust for their hard work in preparing for the review and for their kindness and helpfulness during the course of the visit. We are grateful too, to the users and carers who took time to come and meet the review team. Thanks are also due to the visiting team and their employing organisations for the time and expertise they contributed to this review.

Return to Index

Review Visit Findings

Trust-wide

General Comments

This review looked at the care of health services for children and young people with haemoglobin disorders. During the course of the visit, reviewers met with patients, parents and carers, and with staff providing the services and visited the emergency department, day unit and Ward 3b.

Alder Hey Children's NHS Foundation Trust was reviewed as a Specialist Haemoglobinopathy Team (SHT) for paediatric services and was part of the North West England Haemoglobin Network. As part of the national programme for the reconfiguration of haemoglobinopathy services, the Trust had submitted a joint proposal with the Royal Liverpool University Hospital to become a Haemoglobinopathy Coordinating Centre (HCC) for the care of adults and children with sickle cell disease.

CHILDREN AND YOUNG PEOPLE

Trust	Reviewed as:	No. of children with sickle cell disease	No. of children with thalassaemia	Usually seen by the linked hospitals/local teams (except for annual reviews)	No. of children on long-term red cell transfusions – all
Alder Hey Children's NHS Foundation Trust	SHT	65	10	0	6

Support groups available for patients and carers	Y/N
Sickle Cell Disease	Y
Thalassemia	Y

Specialist Team (Children and Young People Services): Alder Hey Children's NHS Foundation Trust

General Comments and Achievements

This was an experienced service with strong leadership evident throughout. The team were extremely proud of what they had achieved since the last visit in 2015 and it was clear to the reviewers that the team was highly committed and enthusiastic despite there being limited consultant time allocated to care the of patients with haemoglobin disorders. The lead nurses were providing a comprehensive outpatient service with plans to develop transition and community based support and services.

The majority of patients cared for by the team (90%) lived within a small postcode area of Liverpool and close to Alder Hey Hospital. Approximately ten patients were from other hospitals across the regional catchment area for the Trust.

Specialist Haemoglobinopathy Centre	Local Haemoglobinopathy Teams
Alder Hey Children's NHS Foundation Trust	• Aintree University Hospital NHS Foundation Trust • Countess of Chester Hospital NHS Foundation Trust • Bangor Hospital • Glan Clwyd Hospital • Mid Cheshire Hospitals NHS Foundation Trust • Noble's Hospital, Isle of Man • St Helens and Knowsley Teaching Hospitals NHS Trust • Southport and Ormskirk Hospital NHS Trust • Warrington and Halton Hospitals NHS Foundation Trust • Wirral University Teaching Hospital NHS Foundation Trust • Wrexham Maelor Hospital

Staffing

Staffing for the SHC Paediatric Haemoglobinopathy Service [1]	Number of patients	Actual WTE (at time of visit)
Consultant haematologist/paediatrician - >0.6 WTE per 150 patients dedicated to work with patients with HBO	75	0.2
Is at least 0.25 WTE allocated to haemoglobinopathies CPD in the Paediatric Consultant Job	75	Included in SPA time
Trust employ a clinical psychologist for paediatric patients who has >0.5 WTE per 150 patients dedicated to work with patients with HBO ?	75	0.8wte
Trust employ a consultant with a special interest in pain that has 0.1PA of their work plan dedicated to patients with HBO ? Adults/Paeds	75	0

[1] *Recommended Staffing: National NHS England compliance exercise for designation of SHTs 2019.*

Emergency Care

At the time of the visit all patients needing acute admissions were admitted via the children's Emergency Department (ED) and triaged as 'admission stay' or 'short stay' (in which case they could be admitted for up to 24 hours). Patients received analgesia, and cannulation and bloods were performed if necessary. Emergencies were referred to the haematology team and an on-call consultant could be contacted for advice and review. During working hours patients were also seen by one of the two specialist nurses.

Inpatient Care

The new in-patient Ward was co-located with the outpatient clinic and day care facility on Ward 3b. The Ward was a 16 bedded dedicated haematology/oncology unit with twelve rooms with en-suite facilities and one four bedded bay. At the time of the visit the ward was open to 13 beds, with nine rooms with en-suite facilities and one four-bedded bay

The Ward included a dedicated teenage unit and a school room with term-time educational support. There were 42 inpatient admissions during 2018. Occasionally patients were admitted to other Wards within the hospital and were assessed on their priority for transfer to Ward 3b.

Day care

The day care facility had ten beds/ chair spaces and was open from Monday to Friday 8am to 7pm, it was staffed by nurses from the Ward and the two lead nurses. Two bed/chair spaces were also available for day care within the teenage unit. Short stay patients were admitted for day case management of a crisis, and were able to receive intravenous fluids and analgesia before being discharged home or admitted for further treatment if necessary. Patients also attended the unit for pre-transfusion investigations.

Outpatient Care

Outpatient clinics were held in the paediatric haematology/oncology clinic on Ward 3b. A dedicated sickle cell clinic was held on a Thursday afternoon every week and a weekly nurse led clinic on a Wednesday afternoon. A three monthly combined sickle cell/respiratory clinic was due to be reinstated.

Community-based Care

A community nurse based at the hospital had commenced in post. The lead for community care was also the lead for the transition of young people to the adult service based at the Royal Liverpool University Hospital.

The Trust also had plans to develop screening and counselling clinics for those at high risk of being a sickle cell carrier.

Progress since last visit

The previous review had taken place in 2015, and since this time there had been significant progress and a number of improvements:

- The Trust had moved to a new purpose built hospital. The haematology/ oncology teams were now co-located, and staff felt that the merger between the services was working well.
- The hydroxycarbamide programme had been expanded to all patients over nine months of age.
- A weekly nurse-led clinic had been implemented to support the monitoring of patients as well as providing support on the benefits of taking hydroxycarbamide.
- The process for ensuring that children and young people had an annual review had improved. The data showed that 57 out of 65 patients (87%) with sickle cell disease and three out of seven patients (43%) with thalassaemia had an annual review recorded.

- A community nurse specialist had commenced in post and would also be the lead for the care of young people transitioning to adult services.
- A psychologist (0.8wte) with dedicated time allocated for work with patients and carers with haemoglobin disorders had joined the team.
- The Trust was able to provide cardiac T2* magnetic resonance imaging (MRI) scans to improve the monitoring of iron overload.

Views of Service Users and Carers

The visiting team met with four parents during the course of the visit. Overall the parents were highly complimentary about the care their children received and were particularly appreciative of the support from the lead nurses. In addition:

- Parents were grateful that staff had listened to feedback about the change in clinic day and that the clinics were therefore held as before on a Thursday afternoon.
- Most of the parents had children on hydroxycarbamide and they reported that they could access timely support and advice from the lead nurses.
- Concern was expressed that if the lead nurses were not available to perform cannulation then there were often delays, and some carers expressed a lack of confidence in other staff being able to cannulate their child.
- Parents who met with the visiting team reported that they would like more psychology support, and that sometimes requests for support had not been forthcoming.

Good Practice

1. Information leaflets developed by the Trust were excellent. Reviewers were particularly impressed with the following:
    a. A 'crisis diary' for those with sickle cell disease which included prompts for children about each stage of their care so that they could understand more about their condition.
    b. Information for parents and carers about 'non-affected' siblings, which provided useful hints and advice.
    c. Information about medication so that children and young people could understand why they needed to take penicillin and hydroxycarbamide.
    d. Information on alternative pain therapies, which provided well-rounded advice such as including regular exercise.
2. The teaching programme which had been developed to support the transition from paediatric to adult services was well designed. Each young person was given their own 'transition folder', which they could add information to and use as a resource. Space was also included for the young person to document any advice they had been given and any queries that they needed to cover during meetings with the specialist team.
3. An efficient nurse-led hydroxycarbamide service was in operation
4. Targeted education sessions for all girls and boys starting from the age of 11 years, were run throughout the year and to encourage participation some sessions were scheduled during school holidays. The sessions covered all aspects of haemoglobin disorders including diagnosis, management and treatments.

5. The clinical guidelines were well written and were easy to follow, especially for junior staff. Each guideline documented clear information and the steps that staff would need to take. The one page 'acute management pathway' was particularly clear, providing, at a glance, information on how to manage a child experiencing a sickle cell crisis.

6. The pathway for further investigating an abnormal transcranial doppler ultrasound (TCD) was well organised. A dedicated neuroradiologist had oversight of the TCD service and the neuroradiology MRI reporting which helped with imaging of this group.

7. Reviewers were impressed with the level of psychology support dedicated to the care of children and young people with haemoglobin disorders. With the appointment of the dedicated psychologist, patients were able to have a neuro psychology assessment which meant that early help and support could more easily be targeted for those in need. Reviewers were also impressed with overall level of psychological support provided within the Trust for children with long term conditions, especially around key areas of improving emotional health and wellbeing, building resilience and techniques to help manage chronic pain.

8. All patients and their families were given a patient survey questionnaire when they attended their annual review, which ensured that the views of those patients who were seen less frequently were also captured.

Immediate Risks: No immediate risks were identified.

Serious Concern

1. **Consultant Medical Staffing**

   Reviewers were seriously concerned about whether the service had sufficient consultant medical staff for the care of people with haemoglobin disorders to provide staffing for clinics, regular reviews and emergency care. At the time of the visit only one consultant haematologist was available to provide the service, this consultant was providing 24/7 advice on non-malignant haematology and also part of the haematology/ oncology consultant rota (1:5).

   The Trust had two consultant paediatric haematologist vacancies, and approval to appoint to a fourth consultant post. Another colleague was on maternity leave. One locum consultant haematologist was providing some cover, but was on leave for six weeks, and another consultant, who had recently retired, would be returning to provide some short-term support two days a week. Reviewers considered that these arrangements were not sustainable, and highlighted the significant out of hours and 'on service' commitment that the remaining consultant was expected to perform. The haematology service manager and the executive team, who met with the visiting team, were aware of this staffing pressure and had recently met to review the risk to the haematology service. They reported that they were monitoring the situation on a daily basis, had a mitigation plan in place for the next six weeks and were exploring longer term solutions to appoint to the vacant posts. The shortage of consultant staff was documented on the Trust risk register.

Concern

1. **Transcranial Doppler Ultrasound**

   One neuro-radiologist and a sonographer performed all the TCD examinations, but the audit of activity for 2018 showed that, because of the low numbers of patients with haemoglobin disorders cared for by the service, only 60 TCD ultrasounds had been performed. This level of activity did not meet the recommended number of 40 TCD scans being undertaken annually by each practitioner. The team had completed a training competence programme in London some time ago. Although the team has no control over the number of TCD scans it was felt that, in view of the small numbers, there should be a robust internal quality assurance exercise to demonstrate staff competence.

Further Consideration

1. Written information for parents and young people about their long-term condition and also the long-term use of hydroxycarbamide would benefit from review to ensure that sexual health, the possible side effects of long term therapy and fertility, including sperm banking, are clearly explained. Reviewers considered that was particularly important for young people who are transitioning to adult care, to ensure that they have sufficient knowledge to give informed consent.
2. When the lead nurses were absent, ward nurses with competences in cannulation were not available at all times when patients attended the Ward or the day unit for transfusions. Staff who met with reviewers commented that because of the type of patients cared for on the Ward (who tended to have central venous lines in place) and the low numbers of patients with haemoglobin disorders, maintaining staff competences was difficult.
3. Reviewers were shown a number of different combinations of patient records (paper notes, electronic notes and purple mini folders), as the Trust was in the process of implementing an electronic patient care record. Some of the notes that were viewed electronically at the time of the visit did not have the latest version of the patients care plan or annual review letter, as these were documented in hard copy and scanned at a later date. Medical notes were accessible at all times, and reviewers were told that, in practice, staff could access the relevant patient information in an emergency and out of hours. Reviewers suggested that, until the system for electronic patient records was fully implemented the Trust should continue to assure itself that patient safety was not compromised by the use of different formats for recording patient information.
4. A programme of training for ED staff covering haemoglobin disorders was in the process of being re-introduced. Reviewers considered that it would be important to ensure that ongoing training was available, to ensure that staff were aware of the needs of patients with haemoglobin disorders attending the department in an emergency.
5. Some parents who met with the reviewing team commented that their children, who were aged between 14 and 15 years of age, had not yet been commenced on the transition pathway and were unclear about what to expect in terms of transition arrangements.
6. Considering the level of psychology support available, reviewers were unclear why some parents who met with the visiting team felt that the psychology support available was not yet meeting their needs. It may be helpful, once the postholder returns from leave, to work with children and families around their perceptions and expectations of the service and how best to meet their needs.

Return to Index

Commissioning

General Comments and Achievements

The local NHSE commissioning team had no concerns about the service. They felt very positive about the progress that had been made since the last review in 2015. The service had recently submitted a bid to become the Haemoglobinopathy Co-ordinating Centre for sickle cell disease for the North West region and this had the full support of the commissioning team.

Return to Index

APPENDIX 1 Membership of Visiting Team

Clinical Lead		
Mark Velangi	Consultant Paediatric Haematologist	Birmingham Women's and Children's NHS Foundation Trust

Visiting Team		
Nkechi Anyanwu	Clinical Nurse Manager	Evelina London Children's Hospital, Guy's and St Thomas' NHS Foundation Trust.
Louise George	Haematology Nurse Specialist	Sheffield Children's NHS Foundation Trust
Roanna Maharaj	Patient Representative	UK Thalassaemia Society
Janine Younis	Consultant Paediatrician	Whittington Health NHS Trust

QRS Team		
Sarah Broomhead	Assistant Director	Quality Review Service

Return to Index

APPENDIX 2 Compliance with the Quality Standards

Analyses of percentage compliance with the Quality Standards should be viewed with caution as they give the same weight to each of the Quality Standards. Also, the number of Quality Standards applicable to each service varies depending on the nature of the service provided. Percentage compliance also takes no account of 'working towards' a particular Quality Standard. Reviewers often comment that it is better to have a 'No, but', where there is real commitment to achieving a particular standard, than a 'Yes, but', where a 'box has been ticked' but the commitment to implementation is lacking. With these caveats, table 1 summarises the percentage compliance for each of the services reviewed.

Table 1 - Percentage of Quality Standards met

Details of compliance with individual Quality Standards can be found below.

Service	Number of Applicable QS	Number of QS Met	% met
Specialist Haemoglobinopathy Centre – Alder Hey Children's NHS Foundation Trust	50	44	88

Return to Index

Specialist Services for People with Haemoglobin Disorders

Ref	Standard	Met?	Comments
HN-101	**Haemoglobin Disorder Service Information** Written information should be offered to patients and, where appropriate, their carers covering at least: a. Brief description of the service, including times of phlebotomy, transfusion and psychological support services b. Clinic times and how to change an appointment c. Ward usually admitted to and its visiting times d. Staff of the service e. Community services and their contact numbers f. Relevant national organisations and local support groups g. Where to go in an emergency h. How to: i. Contact the service for help and advice, including out of hours ii. Access social services iii. Access benefits and immigration advice iv. Interpreter and advocacy services, PALS, spiritual support and HealthWatch (or equivalent) v. Give feedback on the service, including how to make a complaint vi. Get involved in improving services (QS HN-199)	Y	However, information relating to haemoglobin disorders was not clearly visible in the ward areas visited by the reviewers (information for other conditions was displayed). Some of the commercially produced information (information included was still accurate), was in an older format and reviewers considered that the team may wish to look at obtaining newer versions. Reviewers considered that it would be helpful for patients if information about community services was included now that the team had a CNS who would provide some community support.

Ref	Standard	Met?	Comments
HN-102	**Information about Haemoglobin Disorders** Written information should be offered to patients and, where appropriate, their carers covering at least: a. A description of their condition (SC or T), how it might affect them and treatment available b. Problems, symptoms and signs for which emergency advice should be sought c. How to manage pain at home (SC only) d. Splenic palpation and Trans-Cranial Doppler scanning (children only) e. Transfusion and iron chelation f. Possible complications, including priapism and complications during pregnancy g. Health promotion, including: i. Travel advice ii. Vaccination advice h. National Haemoglobinopathy Registry, its purpose and benefits i. Self-administration of medications and infusions	Y	See also the 'Good Practice' section of the report
HN-103	**Care Plan** All patients should be offered: a. An individual care plan or written summary of their annual review including: i. Information about their condition ii. Plan for management in the Emergency Department iii. Planned acute and long-term management of their condition, including medication iv. Named contact for queries and advice b. A permanent record of consultations at which changes to their care are discussed The care plan and details of any changes should be copied to the patients' GP and their local / specialist team consultant (if applicable).	Y	The template covered all the areas as required by the QS.

Ref	Standard	Met?	Comments
HN-104	**Information for Primary Health Care Team** Written information should be sent to the patient's primary health care team covering available local services and: a. The need for regular prescriptions including penicillin or alternative (SC and splenectomised T) and analgesia (SC) b. Side effects of medication, including chelator agents [SC and T] c. Guidance for GPs on: i. Hydroxycarbamide and iron chelation therapy (if being prescribed by GPs) ii. Immunisations iii. Contraception and sexual health d. Indications and arrangements for seeking advice from the specialist service	Y	The letters seen were very detailed, but reviewers considered that the template letter should be revised to include a more comprehensive section on information about contraception and sexual health (c iii).
HN-105	**Information about Trans-Cranial Doppler Ultrasound (Children's Services Only)** Written information should be offered to patients and their carers covering: a. Reason for the scan and information about the procedure b. Details of where and when the scan will take place and how to change an appointment c. Any side effects d. Informing staff if the child is unwell or has been unwell in the last week e. How, when and by whom results will be communicated	Y	Leaflets for both children, parents and carers were clear about the process for undertaking trans cranial doppler ultrasound examinations.
HN-106	**School Care Plan (Children's Services Only)** A School Care Plan should be agreed for each child or young person covering, at least: a. School attended b. Medication, including arrangements for giving / supervising medication by school staff c. What to do in an emergency whilst in school d. Arrangements for liaison with the school e. Specific health or education need (if any)	Y	

Ref	Standard	Met?	Comments
HN-194	**Environment** The environment and facilities in phlebotomy, out-patient clinics, Wards and day units should be appropriate for the usual number of patients with haemoglobin disorders. Services for children and young people should be provided in a child friendly environment, including toys and books / magazines for children and young people of all ages.	Y	Reviewers were impressed with the environment for children and their families. Each of the rooms had sliding doors which meant that bed and wheelchair access in and out of the rooms was very easy.
HN-195	**Transition to Adult Services** Young people approaching the time when their care will transfer to adult services should be offered: a. The opportunity to discuss the transfer of care at a joint meeting with paediatric and adult services b. A named coordinator for the transfer of care c. A preparation period prior to transfer d. Written information about the transfer of care including arrangements for monitoring during the time immediately afterWards	Y	Each child was given an education work book which covered each stage of the transition pathway. Young people were invited to attend joint meetings with representatives from both the paediatric and adult services. See further consideration section of the report about implementation of the transition pathway.
HN-199	**Involving Patients and Carers** The service's involvement of patients and carers should include: a. Mechanisms for receiving feedback from patients and carers b. An annual patient survey (or equivalent) c. Mechanisms for involving patients and, where appropriate, their carers in decisions about the organisation of the service d. Examples of changes made as a result of feedback and involvement of patients and carers	Y	The parent led support group was active and each year ran sickle cell awareness events. Changes made as a result of feedback include an education programme for those with haemoglobin disorders for age 13yrs and above and a change of consultant outpatient clinic day.
HN-201	**Lead Consultant** A nominated lead consultant with an interest in the care of patients with haemoglobin disorders should have responsibility for guidelines, protocols, training and audit relating to haemoglobin disorders, and overall responsibility for liaison with other services within the network. The lead consultant should undertake Continuing Professional Development of relevance to this role and should have an appropriate number of session/s identified for this role within their job plan and cover for absences should be available.	N	There service had a nominated lead consultant with an interest in patients with haemoglobin disorders but at the time of the visit, due to consultant staff vacancies, had limited time and no cover for lead clinician activities.

Ref	Standard	Met?	Comments
HN-202	**Lead Nurse** A lead nurse should be available with: a. Responsibility, with the lead consultant, for guidelines, protocols, training and audit relating to haemoglobin disorders b. Responsibility for liaison with other services within the network c. Competences in caring for people with haemoglobin disorders d. Competences in the care of children and young people (children's services only) The lead nurse should have appropriate time for their leadership role and cover for absences should be available.	Y	
HN-203	**Medical Staffing and Competences** The service should have sufficient medical staff with appropriate competences in the care of people with haemoglobin disorders, including: **All services:** a. Haematology or paediatric medical staffing for clinics and regular reviews b. 24/7 consultant and junior staffing for emergency care **SHTs only:** c. A consultant specialising in the care of people with haemoglobin disorders on call and available to see patients during normal working hours d. If doctors in training are part of achieving 'a' or 'b' then they should have the opportunity to gain competences in all aspects of the care of people with haemoglobin disorders Staffing levels should be appropriate for the number of patients cared for by the service and its role in the network (SHT/LHT). Staff working with children and young people should have competences in caring for children as well as in haemoglobin disorders. Cover for absences should be available.	N	Medical staffing for the haemoglobin disorder service in an out of hours was reliant one consultant who was undertaking a 1:1 on-call rota due to consultant staff vacancies- see main report.

Ref	Standard	Met?	Comments
HN-204	**Nurse Staffing and Competences** The service should have sufficient nursing staff with appropriate competences in the care of people with haemoglobin disorders, including: a. Clinical nurse specialist/s with responsibility for the acute service b. Clinical nurse specialist/s with responsibility for the community service c. Ward-based nursing staff d. Day unit (or equivalent) nursing staff e. Nurses with competences in cannulation and transfusion available at all times patients attend for transfusion Staffing levels should be appropriate for the number of patients cared for by the service and its role in the network (SHT/LHT). Staff working with children and young people should have competences in caring for children as well as in haemoglobin disorders. Cover for absences should be available.	Y	Only two ward nurses had competences in cannulation (e) as the majority of children admitted to the ward had central venous catheters in place for the administration of intravenous therapies. In practice the lead nurses, Trust IV team and medical staff were available to cannulate children, though parents who met with the visiting team commented about being anxious when staff other than the lead nurses were called to perform cannulations.
HN-205	**Psychology Staffing and Competences** The service should have sufficient psychology staff with appropriate competences in the care of people with haemoglobin disorders, including: a. An appropriate number of regular clinical session/s for work with people with haemoglobin disorders and for liaison with other services about their care b. Time for input to the service's multi-disciplinary discussions and governance activities c. Provision of, or arrangements for liaison with and referral to, neuro-psychology Staffing levels should be appropriate for the number of patients cared for by the service and its role in the network (SHT/LHT). Staff working with children and young people should have competences in caring for children as well as in haemoglobin disorders. Cover for absences should be available.	Y	Since the last visit the Trust had appointed a 0.8 WTE Psychologist dedicated for work with patients and families with haemoglobin disorders. Whilst the psychologist was on maternity leave some cover was available from the lead neuro-psychologist and a trust-wide programme of resilience training was accessible for patients and their families. Parents who met with the visiting team commented that they had felt that the psychology support available was not yet meeting their needs and reviewers considered that the once the postholder returns from leave that further work on how patient expectations could best be met would be beneficial.

Ref	Standard	Met?	Comments
HN-206	**Training Plan** A training plan should ensure that all staff are developing and maintaining appropriate competences for their roles in the care of patients with haemoglobin disorders.	Y	All staff undertook induction training for their work within the haemoglobin disorder service and education meetings were held jointly with the haematology oncology service. Competences for staff had been developed. A practice educator had been appointed who would also ensure that training was delivered on a regular basis.
HN-207	**Trans-Cranial Doppler Ultrasound Competences (Children's Services Only)** Sufficient staff with appropriate competences for Trans-Cranial Doppler ultrasound should be available. Staff should undertake at least 40 scans per annum and complete an annual assessment of competence. Cover for absences should be available.	N	Meeting the recommended 40 TCD examinations annually was not possible due to the population size. One neuro-radiologist and a sonographer did perform all the TCD ultrasound examinations and some joint examinations were undertaken.
HN-299	**Administrative, Clerical and Data Collection Support** Administrative, clerical and data collection support should be appropriate for the number of patients cared for by the service.	Y	
HN-301	**Support Services** Timely access to the following services should be available. In Specialist Centres staff should have specific competences in the care of people with haemoglobin disorders and sufficient time for patient care and for attending multi-disciplinary meetings (HN-602) as required. a. Social worker/ benefits adviser b. Leg ulcer service c. Play specialist (children's services only) d. Chronic pain team (adult services only) e. Dietetics f. Physiotherapy (in-patient and community-based) g. Occupational therapy h. Mental health services (adult and CAMHS) i. DNA studies j. Polysomnography	Y	

Ref	Standard	Met?	Comments
HN-302	**Emergency Department – Staff Competences** Medical and nursing staff working in the Emergency Department/s should have competences in urgent care of people with haemoglobin disorders.	N	A regular training programme for all staff working in the emergency department was in the process of being re-introduced. Nursing staff were invited to attend the ward sessions and the new practice educator had plans to provide department based training for staff.
HN-303	**Specialist On-site Support** Access to the following specialist staff and services should be easily available on the same hospital site as the specialist team: a. Manual exchange transfusion (24/7) b. Erythrocytapheresis c. Acute pain team including specialist monitoring of patients with complex analgesia needs d. High dependency care, including non-invasive ventilation e. Level 2 and 3 critical care	Y	

Ref	Standard	Met?	Comments
HN-304	**Specialist Services - Network** Access to the following specialist staff and services with an appropriate level of specialist expertise in the care of people with haemoglobin disorders should be available: a. Pulmonary hypertension team (adults) b. Consultant obstetrician with an interest in care of people with haemoglobin disorders and specialist high risk anaesthetics (adults) c. Respiratory physician with interest in acute/chronic sickle lung disease and obstructive sleep apnoea (adults & children) d. Fertility, including pre-implantation genetic diagnosis and sperm storage (adults) e. Consultant cardiologist with interest in sickle cardiomyopathy, iron overload related heart disease (adults) f. Consultant endocrinologist with interest in thalassaemia related endocrinopathy and osteoporosis (adults) g. Consultant paediatric endocrinologist with interest in growth problems related to haemoglobinopathies and thalassaemia related endocrinopathy (children) h. Hepatobiliary team with an interest in sickle hepatopathy, viral liver disease, iron overload-related liver disease (adults & children) i. Consultant neurologist and neurosurgeon with an interest in sickle vasculopathy (adults & children) j. Hyperacute stroke service (adults) k. Consultant ophthalmologist with an expertise in sickle retinopathy and chelation related eye disease (adults & children) l. Consultant nephrologist with expertise in sickle nephropathy (adults & children) m. Consultant urologist with expertise in managing priapism and erectile dysfunction (adults & children) n. Orthopaedic service with expertise in managing sickle and thalassaemia related bone disease (adults & children) o. Specialist imaging, including i. MRI tissue iron quantification of the heart and liver ii. Trans-Cranial Doppler ultrasonography (children) p. Bone marrow transplantation services (children only) q. Physiotherapy services (in patient and community based) r. Interventional and neuroradiology for neurovascular complications	Y	
HN-305	**Laboratory Services** UKAS / CPA accredited laboratory services with satisfactory performance in the NEQAS haemoglobinopathy scheme and MHRA compliance for transfusion should be available.	Y	

Ref	Standard	Met?	Comments
HN-401	**Facilities and Equipment** Facilities and equipment should be appropriate for the usual number and case mix of patients. Equipment should be appropriately maintained.	Y	
HN-501	**Transition Guidelines** Guidelines on transition to adult care should be in use covering at least: a. Age guidelines for timing of the transfer b. Involvement of the young person, their carer, paediatric services, primary health care, social care and Local Haemoglobinopathy Team (if applicable) in planning the transfer c. Allocation of a named coordinator for the transfer of care d. A preparation period and education programme relating to transfer to adult care e. Communication of clinical information from paediatric to adult services f. Arrangements for monitoring during the time immediately after transfer to adult care g. Arrangements for communication between the Specialist Haemoglobinopathy Centres and Local Haemoglobinopathy Teams	Y	Good links were in place with representatives from the adult service. Young people were monitored by the paediatric service until they were seen at their first adult clinic appointment, which would also be attended by a member of the paediatric team.
HN-502	**Monitoring Protocols** Protocols should be in use covering the monitoring expected at: a. First out-patient appointment (SHT only) b. Routine monitoring c. Annual review (SHT & any LHTs to which annual reviews are delegated) Protocols should cover both clinical practice and information for patients and families.	Y	The one page guideline for 'crisis' management was very comprehensive and clearly written.
HN-503	**Clinical Guidelines: LHT Management and Referral** Guidelines on routine out-patient monitoring and management between annual reviews should be in use which specify the indications for telephone advice, early referral and immediate transfer to the Specialist Centre.	Y	

Ref	Standard	Met?	Comments
HN-504	**Transfusion Guidelines** Transfusion guidelines should be in use covering: a. Indications for: i. emergency and regular transfusion ii. use of simple or exchange transfusion iii. offering access to automated exchange transfusion to patients on long-term transfusions b. Protocol for carrying out a manual and automated exchange transfusion c. Investigations and vaccinations prior to first transfusion d. Recommended number of cannulation attempts	Y	The advice about the number of cannulation attempts was included in the trust-wide policy.
HN-505	**Chelation Therapy** Guidelines on chelation therapy should be in use covering: a. Indications for chelation therapy b. Choice of chelation drug/s, dosage and dosage adjustment c. Monitoring of haemoglobin levels prior to transfusion d. Management and monitoring of iron overload, including management of chelator side effects e. Use of non-invasive estimation of organ-specific iron overloading heart and liver by T2*/R2 f. Where prescribing is undertaken through shared care arrangements with GPs, guidelines for GPs on prescribing, monitoring and indications for seeking advice from and referral back to the LHT/SHT. g. Self-administration of medications and infusions and encouraging patient and family involvement in monitoring wherever possible.	Y	'f' was not applicable as only the specialist team prescribed medication.

Ref	Standard	Met?	Comments
HN-506	**Clinical Guidelines: Acute Complications** Guidelines on the management of acute complications should be in use covering at least: **For patients with sickle cell disease:** a. Acute pain b. Fever, infection and overwhelming sepsis c. Acute chest syndrome d. Abdominal pain and jaundice e. Acute anaemia f. Stroke and other acute neurological events g. Priapism h. Acute renal failure i. Haematuria j. Acute changes in vision k. Acute splenic sequestration (children only) **For patients with thalassaemia:** l. Fever, infection and overwhelming sepsis m. Cardiac, hepatic or endocrine decompensation	Y	The guidelines were comprehensive but would benefit from revision to include more detail on cardiac, hepatic or endocrine decompensation ('m').
HN-507	**Specialist Management Guidelines** Guidelines should be in use covering the care of patients with sickle cell disease and thalassaemia: a. During anaesthesia and surgery b. Who are pregnant c. Receiving hydroxycarbamide therapy	Y	
HN-508	**Clinical Guidelines: Chronic complications** Guidelines on the management of chronic complications should be in use covering at least: a. Renal disease b. Orthopaedic problems c. Retinopathy d. Cardiological complications / pulmonary hypertension e. Chronic respiratory disease f. Endocrinopathies g. Neurological complications h. Chronic pain i. Liver disease j. Growth delay / delayed puberty (children only) k. Enuresis (children only)	Y	
HN-509	**Referral for Consideration of Bone Marrow Transplantation (Children's Services Only)** Guidelines for referral for consideration of bone marrow transplantation should be in use.	Y	

Ref	Standard	Met?	Comments
HN-510	**Non-Transfusion Dependent Thalassaemia (nTDT)** Network-agreed clinical guidelines for the management of Non-Transfusion Dependent Thalassaemia should be in use, covering: a. Indications for transfusion b. Monitoring iron loading c. Indications for splenectomy	Y	
HN-599	**Clinical Guideline Availability** Clinical guidelines for the monitoring and management of acute and chronic complications should be available and in use in appropriate areas including the Emergency Department, clinic and Ward areas.	Y	

Ref	Standard	Met?	Comments
HN-601	**Service Organisation** A service organisation policy should be in use covering arrangements for: a. 'Fail-safe' arrangements for ensuring all children with significant haemoglobinopathy disorders who have been identified through screening programmes are followed up by a SHT (Children's SHT only) b. Ensuring all patients are reviewed by a senior haematology decision-maker within 14 hours of acute admission c. Patient discussion at multi-disciplinary team meetings (QS HN-602) d. Arrangements for liaison with community paediatricians and with schools (children's services only) e. 'Fail-safe' arrangements for ensuring all children and young people have Trans-Cranial Doppler ultrasound when indicated (children's services only) f. Follow up of patients who do not attend g. Transfer of care of patients who move to another area, including communication with all SHT, LHTs and community services involved with their care before the move and communication and transfer of clinical information to the SHT, LHT and community services who will be taking over their care. h. Accessing specialist advice (QS HN-304) i. Two-way communication of patient information between SHT and LHTs j. If applicable, arrangements for coordination of care across hospital sites where key specialties are not located together	Y	Arrangements for 'fail safe' and management of patients who did not attend appointments were included in other Trust policies.
HN-602	**Multi-Disciplinary Meetings** Multi-disciplinary team meetings to discuss and review patient care should be held regularly, involving at least the lead consultant, lead nurse, nurse specialist or counsellor who provides support for patients in the community, psychology staff and representatives of support services (QS HN-301).	Y	MDT meetings were held weekly and outcomes recorded on the electronic patient record system(Meditech)

Ref	Standard	Met?	Comments
HN-603	**Delegation of Annual Reviews** If annual reviews are undertaken by LHT/s on behalf of the SHT, a written agreement should be in place covering: a. Monitoring protocols (QS HN-502) b. LHT management and referral guidelines (QS HN-503) c. National Haemoglobinopathy Registry data collection (QS HN-701)	N/A	Annual reviews were all undertaken by the specialist team.
HN-604	**Out of Hours Elective Care** Arrangements should be in place for out of hour's transfusion, phlebotomy and out-patient clinics appropriate to the needs of the local population.	Y	
HN-605	**Service Level Agreement with Community Services** A service level agreement for support from community services should be in place covering, at least: a. Role of community service in the care of patients with haemoglobin disorders b. Two-way exchange of information between hospital and community services.	N/A	
HN-606	**Trans-Cranial Doppler Ultrasound Standard Operating Procedure (Children's Services Only)** A Standard Operating Procedure for Trans-Cranial Doppler ultrasound should be in use covering at least: a. Trans-Cranial Doppler modality used b. Identification of ultrasound equipment and maintenance arrangements c. Identification of staff performing Trans-Cranial Doppler ultrasound (QS HN-207) d. Arrangements for ensuring staff performing Trans-Cranial Doppler ultrasound have and maintain competences for this procedure, including action to be taken if a member of staff performs less than 40 scans per year e. Arrangements for recording and storing images and ensuring availability of images for subsequent review f. Reporting format g. Arrangements for documentation and communication of results h. Internal systems to assure quality, accuracy and verification of results	N	The Standard operational procedure did not cover arrangements for 'd' or 'h'.

Ref	Standard	Met?	Comments
HN-607	**Network Review and Learning Meetings** At least one representative of the team should attend each Network Review and Learning Meeting (QS HY-798).	Y	
HN-608	**Neonatal Screening Programme Review Meetings** The SHT should meet at least annually with representatives of the neonatal screening programme to review progress, discuss audit results (QS HN-703), identify issues of mutual concern and agree action.	Y	
HN-701	**National Haemoglobinopathy Registry** Data on all patients, following patient or parental consent, should be entered into the National Haemoglobinopathy Registry. Data should include annual updates, serious adverse events, pregnancies and patients lost to follow up.	Y	Sixty-five out of a possible eighty-five patients had consented for their data to be included on the NHR.
HN-702	**Activity Data** The service should monitor on an annual basis: a. Number of acute admissions, day unit admissions, Emergency Department attendances and out-patient attendances b. Length of in-patient stays c. Re-admission rate d. 'Did not attend' rate for out-patient appointments	Y	

Ref	Standard	Met?	Comments
HN-703	**Quality Dashboard** The service should monitor Quality Dashboard data including the number and percentage of: a. Adverse events reported on the NHR for which a mortality or serious case review has taken place b. Children who have had Trans-Cranial Doppler screening undertaken within national guidelines c. Patients given pain relief within half an hour of presentation with sickle crisis d. Patients with possible sickle disorders identified by neonatal screening who have been entered on a care pathway e. Eligible children beginning penicillin at or before three months of age f. Patients registered on the National Haemoglobinopathy Registry who had an annual review undertaken within the last year g. Patients on long-term transfusion who received cardiac MRI, and the proportion of those receiving a cardiac MRI who achieved a figure of less than 20ms h. Eligible patients with sickle cell disease who received an MRI for liver iron, and the proportion of those who received an MRI for liver iron who achieved more than 7 mg/gm/DW (sickle cell and thalassaemia separately)	Y	
HN-704	**Other Quality Data** The service should monitor on an annual basis: a. Proportion of children who have been offered treatment following Trans-Cranial Doppler Screening	Y	
HN-705	**Other Audits** Clinical audits covering the following areas should have been undertaken within the last two years: a. Availability of extended red cell phenotype in all patients and the proportion of patients who have developed antibodies b. Whether all eligible patients on long term transfusion have been offered automated exchange transfusion c. Waiting times for elective: i. Phlebotomy ii. Cannulation iii. Setting up of the blood transfusion (for pre-ordered blood)	Y	

Ref	Standard	Met?	Comments
HN-706	**Network Audits** The service should participate in agreed network-wide audits.	Y	The network audit programme included audits covering; time to analgesia, immunisation rates and use of hydroxycarbamide.
HN-707	**Research** The SHT should actively participate in research relating to the care of patients with haemoglobin disorders.	Y	
HN-708	**Trans-Cranial Doppler Quality Assurance (Children's Services Only)** The service should monitor and review at least annually: a. Whether all staff performing Trans-Cranial Doppler ultrasound have undertaken 40 procedures in the last year (QS HN-207) b. Results of internal quality assurance systems (QS HN-606) c. Results of National Quality Assurance Scheme (NQAS) for Trans-Cranial Doppler Ultrasound (when established) or local peer review arrangements (until NQAS established)	N	The audit for 2018 showed that neither practitioner met the required number of procedures per year (40). Sixty TCD ultrasounds had been performed of which 33 had been undertaken by the neuroradiologist and 25 by the sonographer. Two examinations had been reported by both practitioners.
HN-798	**Review and Learning** The service should have appropriate multi-disciplinary arrangements for review of, and implementing learning from, positive feedback, complaints, outcomes, audit results, incidents and 'near misses'. This should include: a. Review of any patient with a serious adverse event or who died b. Review of any patients requiring admission to a critical care facility c. Annual review of trends in National Haemoglobinopathy Registry data, activity data, Quality Dashboard, other quality data and other audits (QSs HN-701 to HN-705)	Y	
HN-799	**Document Control** All policies, procedures and guidelines should comply with Trust (or equivalent) document control procedures.	Y	

Return to Index