Health services for people with haemoglobin disorders

University Hospitals Bristol NHS Foundation Trust – Adult Services

Visit Date: 13th November 2019

Report Date: February 2020

8831

Contents

Introduction

This report presents the findings of the peer review of health services for people with haemoglobin disorders in University Hospitals Bristol NHS Foundation Trust that took place on 13th November 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 during the course of the review visit reviewers did enquire about the local network and commissioning arrangements.

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 University Hospitals Bristol 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:

- University Hospitals Bristol NHS Foundation Trust
- NHS England & NHS Improvement Specialised Commissioning – Haemoglobinopathies
- NHS Bristol Clinical Commissioning Group

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 for 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 University Hospitals Bristol 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 also grateful to the users and carers who took the 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

Since the last peer review visit in 2015 there had been a number of changes to the commissioning of the service provided by University Hospitals Bristol NHS Foundation Trust (UHBFT). Despite discussions with the specialist commissioners, the service had been operating as a Local Haemoglobinopathy Team (LHT) because of issues with reimbursement. Following national compliance and procurement exercises conducted by NHS England (NHSE), the Trust had been formally recognised in July 2019 as a Specialist Haemoglobinopathy Team (SHT) for adults and children, and in October 2019 it had been designated as a Haemoglobinopathy Coordinating Centre (HCC) for Sickle Cell Disease. The Trust was keen to support this development and were working with the team to implement the action plan agreed with NHSE. At the time of the visit, discussions were ongoing with NHSE specialist commissioners to agree funding for additional support. Once in operation the SHT would link with the North West London and North Central London HCC for Thalassemia.

The adult haemoglobinopathy service was responsible for about 109 adult haemoglobinopathy patients (85% sickle cell disease and 15% thalassaemia).

As part of the work to develop a specialist service across the south west of England, approximately 50 patients with sickle cell disease had been identified as being cared for at other hospitals across the south west of England, and work was in progress to identify the number of patients across the network with thalassaemia and rare anaemias.

ADULTS

Trust	Reviewed as:	No. of adults with sickle cell disease	No. of adults with thalassaemia	No. of adults on long-term red cell transfusions
University Hospitals Bristol NHS Foundation Trust	SHT	90	19	13

Support Groups	
Sickle Cell Disease – Adults	0
Thalassaemia – Adults	0

Specialist Team (Adult Services): University Hospitals Bristol NHS Foundation Trust

General Comments and Achievements

This was an enthusiastic team, keen to develop a comprehensive SHT service across the network. There was evidence of collaborative working and good relationships with the managerial team.

The haemoglobinopathy team consisted of two consultant haematologists (0.325 wte). The lead clinician was also the lead for the transfusion service. In addition to haemoglobinopathy work, the deputy lead's time included work with patients with lymphoma and those undergoing CART-cell therapy.

The CNS team (1.6 wte) provided advice to members of the primary health care team and also liaised with the antenatal screening service. The CNSs had good links with the paediatric service, providing support for young people transitioning into adult care. The CNS team also had plans to develop nurse-led clinics in the future.

Some psychology support (0.1 wte) was available for patients with haemoglobin disorders. Referral to the psychologist was via the multi-disciplinary team (MDT).

An in-hospital apheresis unit was operational and provided an elective and emergency red cell exchange service.

The SHT had a strong commitment to training and education, with regular teaching delivered by the new consultant and the CNS team to a range of staff groups within the Trust and across the local health economy.

The team were actively engaged with a phase II clinical trial aiming to determine the safety of a PDE9 inhibitor (IMARA), and had expressed an interest in opening a similar trial for patients with thalassaemia.

Specialist Haemoglobinopathy Team	Trusts linked to the SHT
University Hospitals Bristol NHS Foundation Trust	• Gloucestershire Hospitals NHS Foundation Trust • North Bristol NHS Trust • Northern Devon Healthcare NHS Trust • Royal Cornwall Hospitals NHS Trust • Royal Devon and Exeter NHS Foundation Trust • Royal United Hospitals Bath NHS Foundation Trust • Taunton and Somerset NHS Foundation Trust • Torbay and South Devon Healthcare NHS Trust • University Hospitals Plymouth NHS Trust • Weston Area Health NHS Trust • Yeovil District Hospital NHS Foundation Trust

Staffing

Staffing for the Adult Specialist Haemoglobinopathy Service (SHT)[1]	Number of patients	Actual wte (at time of the visit)	NHSE recommended staffing wte
Consultant haematologist with >0.6 wte per 150 patients dedicated to work with patients with haemoglobinopathies	109	0.325	0.44
At least 0.25 wte allocated to haemoglobinopathies CPD in the adult consultant Job plan	109	As part of general CPD allocation	-

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

Staffing for the Adult Specialist Haemoglobinopathy Service (SHT)[1]	Number of patients	Actual wte (at time of the visit)	NHSE recommended staffing wte
A clinical psychologist for adult patients with >0.5 wte per 200 patients dedicated to work with patients with haemoglobinopathies	109	0.1	0.27

Emergency Care

Patients had a contact card and would contact either the CNSs within working hours or the day unit during opening times. Out of hours they were advised to phone the 24-hour emergency line. Patients were either asked to attend the day unit, where they were reviewed by a member of the SHT, or, out of hours, the nurse in charge of the ward would contact the on-call SHO who would then contact the on-call Specialist Registrar (SpR) who would assess the patient.

If there were no beds available on the haematology ward, patients were asked to attend the Emergency Department (ED) where they were reviewed by the attending haematologist. Patients who came directly to the ED were not usually known by the team.

In-patient Care

The haematology department was a 24-bedded unit that cared for patients with non-malignant and malignant haematology conditions.

Day Care

The haematology day unit was open from 8am to 6pm (Monday to Friday) and was open on Sundays from 9am to 5pm. Patients were advised to contact the service on the designated telephone number when they were unwell, and could attend the day unit when the day unit was in operation. There was also a designated 24-hour telephone line to the ward. The apheresis unit was located in the Haematology & Oncology Centre and was open Monday to Friday, 8am to 5pm, for elective and emergency red cell exchanges; it provided an on-call service.

Community-based Care

There was no dedicated community-based nurse for patients with haemoglobin disorders. The hospital based CNSs did provide information for GPs, practice nurses and midwives to help care for patients, and there was a dedicated telephone line that patients and professionals could use to make contact with the team. The Bristol Organisation for Sickle Cell Anaemia Research and Thalassaemia support group (OSCAR) was no longer in operation because of the loss of local funding.

Progress since Last Visit

- Specialist Haemoglobinopathy Team (SHT) status had been granted (as of July 2019) following a successful application. At the time of the visit the team were working with NHSE to implement an action plan, which included additional community support.
- The Trust had also been granted Haemoglobinopathy Coordinating Centre (HCC) status for sickle cell disease for the south west of England in October 2019.
- A programme of teaching for staff had been implemented across the Trust.
- An additional CNS (0.6 wte) had been appointed in January 2019.
- An additional consultant haematologist had been appointed who had two programmed activities (PA) allocated for work with haemoglobinopathies.

Views of Service Users and Carers

The visiting team spoke to two patients via separate telephone calls during the course of the visit, and viewed nine patient feedback questionnaire responses. The feedback may therefore not be representative of other service users who were cared for by the team at UHBFT.

Overall, the two patients who spoke to the reviewers were complimentary about the team. In addition:-

- Access to the CNS worked well and the patients knew how to contact the team.
- Appointments were planned and clinics ran well.
- Both the patients received copies of their letters to GPs following consultations with the SHT.
- CNSs coordinated appointments around patients' work.
- The CNS team were 'brilliant' and 'outstanding'. Advice was accessible for 'anything', including coordination of clinical appointments, advice and documentation for travel, how to access psychology support and access to a social worker.
- Both the patients appreciated the process whereby they received text messages to remind them of upcoming appointments.
- Education had been made available on how to manage pain and when to seek further advice.
- Access to medications worked well; medication was usually prescribed via patients' GPs.

Good Practice

1. The process in place to communicate with primary care and other organisations was very good. The team had arrangements in place with the community and local university medical providers who would alert the team when new patients moved into the area, and the team would contact the students to offer ongoing care locally during term time. The information contained in the letters to the primary health care team was also well written and included clear guidance for GPs.
2. Reviewers were impressed with the work of the specialist nursing team, who were knowledgeable, committed and had clear ownership of the service they provided. The patients who spoke to the reviewers also spoke highly of the CNS team.
3. The annual review proformas in use for patients with sickle cell disease and thalassaemia were very well designed and clearly stated who to contact for queries and advice.
4. The day unit was open on Sundays from 9am to 5pm and provided access for patients who required an emergency assessment or pain management.
5. The service organisation policy was well written, included clear advice and was regularly updated.
6. Good arrangements were in place for the local multi-disciplinary team (MDT) meetings. The MDT meetings were held weekly and the discussion of patients was subdivided into those who had attended clinics, those who had attended the day unit and in-patients. Time was also allocated for general communication and information sharing.

Immediate Risk

1. **Chelation guidelines**[2]

Two versions of the chelation policy were in circulation; the **first**, a specific document for chelation updated in 2019, and the **second** contained in the guideline pertaining to the general management of sickle cell disease. The former had incorrect doses for the initiation and continuation of deferasirox treatment, and referred to the doses applicable to the old formulation (soluble) which was withdrawn at least two years earlier. The higher dose of the soluble tablets could have led to toxicity (renal or hepatic), particularly in patients with poor reserve in this area.

Serious Concern

1. **Specialist expertise**

At the time of the last visit in 2015, the team was reviewed as a specialist haemoglobinopathy team (SHT) in derogation from NHSE, as it was only providing a certain level of specialist work. The Trust had since submitted 'external revenue proposals' annually to NHSE which had not been successful, and it had therefore made the decision to provide an LHT function only, until officially commissioned as an SHT in July 2019. It was unclear whether the commissioners had agreed with this decision. Reviewers were seriously concerned that the Trust team, in a relatively short time, were expected to 'step up' from being an LHT to providing a comprehensive SHT service, and to provide an HCC for people with sickle cell disease. The following reasons lay behind the reviewers' concerns: -

a. The team, until July 2019, had been functioning as an LHT, and working in isolation without any links to an SHT for advice and support, the multi-disciplinary discussion of patients with complex needs or formal arrangements for shared care of patients with haemoglobin disorders.

b. The team had limited experience with which to provide a specialist service and oversight for the region, as it had only been designated as an SHT in July 2019 and HCC for sickle cell disease in October 2019.

c. The team had not been linked to a haemoglobin disorder network; such a network would have provided the LHT at UHBFT with opportunities for education and training, access to clinical guidelines, and network-wide review and learning.

d. The SHT would be providing a specialist service locally and for the south west of England, both of which are low prevalence areas for patients with haemoglobin disorders. The team cared for only 109 patients, and an initial scoping exercise undertaken by the team had identified only 50 additional patients with haemoglobin disorders who were being cared for at local Trusts and who were not yet linked to the SHT. A regional MDT was not yet in place.

e. Reviewers were not assured that there were robust arrangements in place to access staff and services in other specialities (e.g. renal medicine) with an appropriate level of specialist expertise in the care of people with haemoglobin disorders. Speciality services were available at UHBFT; however, reviewers were told that there had only been one referral for surgery and one referral to the renal team in the last two years.

---

[2] **Trust response:** ***Completed actions****: 1. Report as an incident as part of clinical governance processes – upload onto Datix system to follow Trust process. 2. Removed Iron Chelation Guidelines from the document management system 3. Iron Chelation guidelines reviewed by Pharmacy and Medical teams. 4.Updated guideline signed off by the BHOC Clinical Governance Committee 5. Confirmation of which other Centres had been sent the guidelines for use. 6. Removed UHB Iron Chelation Guidelines from use in any other centres. 7.* ***Audit completed in December 2019*** *to determine if there had been any patient harm as a result of the use of the incorrect guideline. A retrospective review was undertaken with pharmacy to assure every prescription of Exjade was screened in pharmacy for the formulation and dose dispensed. Audit results confirmed that all patients were on the correct chelation dose.*
QRS Response to Trust action: *The Trust actions to address the immediate risk were structured, detailing each step clearly. The actions as described will address the immediate risk identified during the visit, once fully completed.*

Reviewers suggested that during the development of the service the SHT should link with another SHT/HCC to ensure that lead staff attain sufficient professional development and specialist expertise in the care of people with haemoglobin disorders. Engagement with the national panel for haemoglobin disorders, once in operation, will also provide additional support to the SHT/HCC for patients with more complex needs.

Concerns

1. **Consultant staffing**

   The service had insufficient consultant medical staff with appropriate competences in the care of people with haemoglobin disorders to provide staffing for regular reviews, emergency care, clinics, and support and oversight to the LHTs across the network. At the time of the visit there were two consultant haematologists (0.125 wte and 0.2 wte respectively) for the adult haemoglobinopathy service. In practice, the deputy lead for the service was seeing patients at any time when on duty. The lead clinician did not provide evidence of any recent relevant CPD for their work with haemoglobin disorders. The designation in October 2019 of the service as an HCC would also require an increase in the time available for leadership and support to other adult haemoglobinopathy services across the region.

2. **Thalassaemia guidance**

   Guidelines covering the acute management of fever, acute symptoms, and care when pregnant for patients with thalassaemia were not in place.

3. **Clinical decision making**

   In three out of the five sets of care records seen at the time of the visit, the rationale behind some clinical decisions was not clear, as the care documented did not appear to follow the clinical guidelines. Reviewers considered that undertaking a wider review of case notes may be helpful to identify if this was due to poor record keeping or the lack of implementation of the agreed clinical guidelines.

4. **Competences and training plan**

   A competence framework was not yet in place for staff on those wards to which people with haemoglobin disorders were usually admitted, to identify whether staff had appropriate competences to care for patients with haemoglobin disorders.

   A training plan showing that all staff were developing and maintaining appropriate competences for their roles in the care of patients with haemoglobin disorders was also not in place. The SHT did provide a programme of training sessions for staff.

5. **Community-based care**

   A community service to provide support for patients and their carers was not commissioned. Reviewers were told that, as part of the development of the SHT, consideration was being given to increasing the level of support to patients in the community by the development of a community support worker role. The hospital-based CNSs did provide a telephone advice service.

6. **Patient information**

   The patient information seen by the reviewers covering the use of deferasirox and hydroxycarbamide was out of date and did not include more recent guidance. Reviewers were told that more up to date information on the use of deferasirox was available to patients in the clinical areas.

Further Consideration

1. Reviewers noted that the undated audit provided showed that only 26% of patients had extended red cell phenotype. However, in addition a further 72% of patients were genotyped. An audit of the proportion of patients who had developed antibodies was planned.

2. The psychology provision was below the recommended staffing levels for the number of patients actively cared for by the service. At the time of the visit, the psychologist had 0.1 wte for work with patients with haemoglobin disorders, and could only see two patients per week. Referral to the service was via the MDT. The psychologist did not have any time to provide other support such as education and group work. As part of the SHT development, there were plans to increase the psychology staffing time available to 0.3 wte in 2020.

3. The Trust teenage and young person's unit did not accept teenagers and young people with haemoglobin disorders. Depending on their age, young people were admitted to either the paediatric or the adult ward.

4. Feedback from patients was limited, and staff who met the reviewing team had plans to repeat the patient survey. Only 9 out of 15 patients had responded to the previous patient survey, and feedback from patients was in contrast to the views of the two patients who spoke to the reviewing team during the course of the visit.

5. The process for reviewing data on the National Haemoglobinopathy Registry (NHR) and quality dashboard would benefit from being formalised to ensure that actions following data review are outcome directed.

6. Reviewers spoke to a representative from one referring hospital who considered that the support from the team was helpful but rarely had a need to contact the team. The referring team would value more formalised support and access to education on haemoglobin disorders. The referring team did not have access to a formal MDT, and no process for annual reviews was yet in place.

Return to Index

Commissioning

The report contains a number of issues which would need to be addressed in order for the adult team to function as a comprehensive Specialist Haemoglobinopathy Team (SHT) and Haemoglobinopathy Coordinating Centre (HCC) for the south west of England.

Immediate Risk: see Trust-wide section of the report.

Serious Concern: see Trust-wide section of the report.

Concern

1. **Access to specialist care**

   Commissioning arrangements, including specialist and co-commissioned services, had not progressed since the last visit in 2015. Reviewers were concerned that patients living in the south west of England had not had access to specialist care – for example, those with complex needs were not being discussed within the wider context of a specialist MDT, and annual reviews were not being completed – and that this had the potential for patients to receive suboptimal treatment.

Return to Index

APPENDIX 1 Membership of Visiting Team

Clinical Lead		
Dr Emma Drasar	Consultant Haematologist	Whittington Health NHS Trust

Visiting Team		
Doris Dennis	Haemoglobinopathy Specialist Nurse / Counsellor	Manchester University NHS Foundation Trust
Prof. Jo Howard	Consultant Haematologist / Honorary Professor in Haemoglobinopathies	Guy's and St Thomas' NHS Foundation Trust
Judith St Hilaire	CNS Haemoglobinopathies / Iron Chelation	Imperial College Healthcare NHS Trust
Dr Ryan Mullally	Consultant Haematologist	Nottingham University Hospitals 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
University Hospitals Bristol NHS Foundation - Adult Services	44	26	59
Total	**44**	**26**	**59**

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	
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	The patient leaflet covering the use of hydroxyurea would benefit from being updated. The information on iron chelation (e) seen by the review team was also out of date, though reviewers were assured by the team that patients were given accurate written information.

Ref	Standard	Met?	Comments
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	From the care records seen, the process for annual reviews was very comprehensive.
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	Information sent to the primary health care team was well written and included clear guidance for GPs.
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	N/A	

Ref	Standard	Met?	Comments
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)	N/A	
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	
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	N	Written information for young people about the transfer of care, including arrangements for monitoring during the time immediately afterwards, was not available. Young people transitioning to the adult service did have the opportunity to meet with representatives from the adult team.
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 patient survey sample response rate was low. Only 9 responses were received from the 15 patients contacted. Reviewers considered that the team should try to gain patient views on access to appropriate analgesia in any future survey.

Ref	Standard	Met?	Comments
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	The lead consultant had insufficient time in their job plan (1.25 PA) for leadership of the SHT and liaison with LHTs across the network. The lead consultant was also the lead for the blood transfusion service.
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 **SHCs 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 (SHC/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	Haematology staffing was insufficient for clinics, regular reviews and the team's role in the network. The lead clinician and deputy had a total of 3.25 PAs allocated for the care of patients with haemoglobin disorders. Reviewers considered that a minimum of 4.4 PAs would be required for the SHT. There was no evidence that the lead clinician had completed any recent CPD for haemoglobin disorders. The deputy lead had completed relevant CPD.

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 (SHC/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	A competence framework for ward and day unit staff was not in place. An education programme was provided by the CNS. Staff had competences in delivering blood transfusions, and had skill in the cannulation of patients.
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 (SHC/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	The psychology provision was below the recommended staffing levels for the number of patients actively cared for by the service. At the time of the visit the psychologist had 0.1 wte for work with patients with haemoglobin disorders, and could only see two patients per week. Referral to the service was via the MDT. The psychologist did not have any time to provide other support such as education and group work. As part of the SHT development there were plans to increase the psychology staffing time available to 0.3 wte in 2020.The suggested workforce recommendation of the British Psychology Society Special Interest Group for Psychologists working in Sickle cell and Thalassaemia (2017) is 1 wte: 300 patients.

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.	N	A training plan showing that staff were developing and maintaining appropriate competences for their roles in the care of patients with haemoglobin disorders was not in place. Training sessions for staff did take place.
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/A	
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.	N	The administrative post was vacant at the time of the review and there was no support for data collection.
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	
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.	Y	Patients with acute pain were asked to attend the day unit (Monday to Friday, 9am to 5pm). Outside these times the on-call medical staff were contacted to see patients on the ward.

Ref	Standard	Met?	Comments
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	N	Whilst specialist services were available, reviewers considered that the low number of patients referred for specialist advice (reviewers were told of two patients in the last two years) meant that the speciality teams would have inadequate expertise in the care of people with haemoglobin disorders. See main report.
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	
HN-502	**Monitoring Protocols** Protocols should be in use covering the monitoring expected at: a. First out-patient appointment (SHC only) b. Routine monitoring c. Annual review (SHC & any LHTs to which annual reviews are delegated) Protocols should cover both clinical practice and information for patients and families.	Y	The template used for completion of annual reviews was well designed.
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.	N/A	This QS is not applicable for SHTs.

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	However, a number of typographical errors were noted and communicated to staff to amend.
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/SHC. g. Self-administration of medications and infusions and encouraging patient and family involvement in monitoring wherever possible.	N	See Immediate Risk section in the main report.
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	N	Clinical guidelines for acute management for patients with thalassaemia ((l) and (m)) were not in place. All other aspects of the QS were met.

Ref	Standard	Met?	Comments
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	N	Guidance covering the care of patients with sickle cell disease and thalassaemia care when pregnant did not include any pre-pregnancy optimisation advice, and did not cover the care of patients who were transfusion dependant.
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)	N	Clinical guidelines covering chronic complications for patients with thalassaemia were not in place.
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.	N/A	
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 SHC (Children's SHC 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 SHC, LHTs and community services involved with their care before the move and communication and transfer of clinical information to the SHC, 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 SHC and LHTs j. If applicable, arrangements for coordination of care across hospital sites where key specialties are not located together	Y	The operational policy was updated regularly and included a good proforma for the follow up of patients who 'did not attend'. The information about the transfer of care to other services was brief and there would be a benefit in expanding this with more detail about the arrangements.
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	Multi-disciplinary team meetings were well attended. Reviewers were impressed with the minutes seen, which were structured in the same way each week to cover the discussions that took place, for example, clinics, in-patients and the communication of any updates.
HN-603	**Delegation of Annual Reviews** If annual reviews are undertaken by LHT/s on behalf of the SHC, 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	The team had not been functioning as an SHT prior to July 2019 but had started to identify patients across the network.

Ref	Standard	Met?	Comments
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	No service level agreement with any community services was in place. The team did have informal arrangements for liaison with midwives for advice.
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/A	
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).	N	Network review and learning meetings were not in place.
HN-608	**Neonatal Screening Programme Review Meetings** The SHC 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	

Ref	Standard	Met?	Comments
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.	N	The team were in the process of submitting data on patients, although there was insufficient administrative support available to support data collection. Not all patients had given consent for their data to be submitted onto the NHR, and reviewers considered that further work to explain the rationale to patients may be helpful.
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	
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)	N	Data had been collated but had not been submitted by the Trust.

Ref	Standard	Met?	Comments
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	N/A	
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	However, see main report on the percentage of patients who had had their red cell phenotype completed.
HN-706	**Network Audits** The service should participate in agreed network-wide audits.	N	A network was not in operation.
HN-707	**Research** The SHC 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/A	

Ref	Standard	Met?	Comments
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	Form the minutes seen, the review of annual review trends and other quality data was not clear. Reviewers commented that the process for the review of data could be more robust, and the actions following review developed to be more outcome directed.
HN-799	**Document Control** All policies, procedures and guidelines should comply with Trust (or equivalent) document control procedures.	Y	

Return to Index