Health Services for People with Haemoglobin Disorders

Sheffield Children's NHS Foundation Trust

Visit Date: 16th October 2019

Report Date: January 2020

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Introduction

This report presents the findings of the peer review of health services for people with haemoglobin disorders in Sheffield Children's NHS Foundation Trust that took place on 16th October 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 Sheffield 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:

- Sheffield Children's NHS Foundation Trust
- NHS England & NHS Improvement Lead Commissioner for Blood and Infection
- NHS England & NHS Improvement Regional Specialised Commissioner

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 Blood and Infection.

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 Sheffield 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 also grateful 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.

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Review Visit Findings

Trust-wide

General comments

This review looked at the health services provided for children and young people with haemoglobin disorders at Sheffield Children's NHS Foundation Trust (SCH). During the course of the visit, reviewers visited the emergency department, paediatric ambulatory care facilities, day units, inpatient wards and the paediatric outpatient department, and met with patients and carers and with staff providing the services for the local health economy.

Sheffield Children's Hospital provided acute and tertiary specialist services for 2.5 million children residing across Yorkshire and Humberside.

CHILDREN AND YOUNG PEOPLE

Trust	Reviewed as:	No. of children with sickle cell disease	No. of children with thalassaemia	No. of children on long-term red cell transfusions
Sheffield Children's NHS Foundation Trust	SHT	62	8	8

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

General Comments and Achievements

This was a small team with strong medical and nursing 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 worked well together and were highly committed and enthusiastic in their vision to provide a high quality service.

The team consisted of the lead consultant, who had joined the team in 2018, two clinical nurse specialists (CNS) (total 1.4 whole time equivalent including 0.4 wte for haemoglobinopathy and 1.0 wte for benign haematology including haemophilia), and a data manager (5 hours per week). A nurse counsellor, who was community-based, liaised with new-born screening and would make initial contact with families.

Approximately 70 children and young people from Sheffield and the surrounding areas were cared for by the team. Representatives from local referring teams and families who took part in discussions during the course of the visit all considered that they had good relationships with the SHT and received timely information after any clinic attendances for their records/care plans.

The team at SCH had good collaborative working relationships with the adult SHT at Sheffield Teaching Hospitals NHS Foundation Trust. Monthly multidisciplinary team (MDT) meetings were held, with a joint MDT meeting held every two months with the adult SHT to discuss complex patients and young people who were in the process of transitioning or had already transitioned to adult care.

In October 2019, following the national procurement exercise conducted by NHS England (NHSE), the service, in conjunction with the adult service at Sheffield Teaching Hospitals NHS Trust, had been formally designated as a Haemoglobinopathy Coordinating Centre (HCC) for Sickle Cell Disease for the north east and Yorkshire. The Trust was keen to support this development and was 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.

Overall, reviewers were impressed with team, who were providing a very good clinical service for children and young people with haemoglobin disorders within considerable staffing constraints.

Specialist Haemoglobinopathy Team	Local Haemoglobinopathy Teams
Sheffield Children's NHS Foundation Trust	• Barnsley Hospital NHS Foundation Trust • Chesterfield Royal Hospital NHS Foundation Trust • Doncaster and Bassetlaw Teaching Hospitals NHS Foundation Trust • Northern Lincolnshire and Goole NHS Foundation Trust (Grimsby, Scunthorpe, Goole) • The Rotherham NHS Foundation Trust • United Lincolnshire Hospitals NHS Trust (Lincoln County Hospital and Pilgrim Hospital Boston)

Staffing

Staffing for the SHC Paediatric Haemoglobinopathy Service [1]	Number of patients	Actual WTE (at time of visit)	Staffing required as recommended by NHSE
Consultant haematologist/paediatrician with >0.6 wte per 150 patients dedicated to work with patients with haemoglobinopathies	70	0.2	0.28
At least 0.25 wte allocated to haemoglobinopathies CPD in the paediatric consultant job	70	None	-
A clinical psychologist for paediatric patients with >0.5 wte per 150 patients dedicated to work with patients with haemoglobinopathies	70	0	0.23

Emergency care

All children and young people with haemoglobin disorders had direct access to the haematology/oncology ward. The emergency care pathway for children and young people had been revised following the last 'time to analgesia' audit, which had identified large differences in the timeliness of care provided to children presenting via the Emergency Department (ED) and the ward. The pathway had been changed so that parents or carers could call the ward directly if a child was unwell and a decision would then be made (depending on whether the haematology medical staff would be immediately available) about where best to see the child for a clinical review. During working hours children would normally be seen in the haematology day care unit. Out of hours, parents and carers were asked to bring the child either to Ward 6 (which had one triage bed) or to the ED, where they would be triaged as urgent, if it was likely that there would be a delay in assessing the child on the ward.

Children and young people from the surrounding areas would access their local hospital for emergency care. The local hospitals had access to shared care guidelines that, as well as providing immediate practical guidance, asked the paediatric team to contact the haematology team at Sheffield Children's Hospital to discuss any patients with haemoglobin disorders.

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

In-patient care

Ward 6 was the dedicated paediatric haematology/oncology ward and had 14 in-patient beds (eight cubicles and six beds in bays). A dedicated school room and a play room with play specialists were located in close proximity to the ward, to allow children to attend school whilst undergoing treatment. An acute pain team provided clinical oversight for all patients on patient-controlled analgesia (PCA), and provided consultative support to patients with complex pain issues.

Day care

Patients were seen in a purpose-built out-patient and day care department for the exclusive use of haematology/oncology patients. There was a large waiting area with toys and activities available, and an adolescent area. The unit was open Monday to Friday between the hours of 8am and 6pm. The large day care unit was able to provide blood sampling, exchange transfusions and top-up transfusions. The unit had a mixture of beds and reclining chairs.

Out-patient care

A dedicated haematology/oncology clinic, with blood-taking and counselling facilities, was located adjacent to the day unit area and was used for routine clinics and unplanned reviews. A weekly haemoglobinopathy clinic was held each Thursday morning, and patients could also be seen in the general haematology clinic held on Wednesday afternoons.

Community-based care

Paediatric community-based care was limited. Some care was provided by community nursing teams who would undertake some blood testing, and by the haemoglobinopathy nurse counsellor (0.4 wte), who also provided support for the adult SHT based at Sheffield Teaching Hospitals NHS Foundation Trust.

The SCH haemoglobinopathy CNS would also provide some support to patients in the community, and attended some schools to provide support with school care plans and training, when capacity allowed. The CNS had well-established communication links with schools that she was unable to visit. Community support by the SCH team was not provided to children who lived outside Sheffield, although contact would be made with these children when they were admitted to SCH or attended clinics there.

Progress since last visit:

- The service had moved to a new dedicated haematology/out-patient area for haematology/oncology
- The service had agreed a service level agreement with NHS Blood & Transplant to provide 24/7 automated exchange transfusions
- The team was participating in the STAND (crizanlizumab) research study
- A data manager had been recruited
- The level of data submitted to the national haemoglobinopathy registry (NHR) had increased since the appointment of the data manager
- Sheffield Teaching Hospitals NHS Trust had been formally recognised as a Haemoglobinopathy Coordinating Centre (HCC) for Sickle Cell Disease and, as SCH was part of the SHT, SCH would be working in collaboration with Sheffield Teaching Hospital NHS Trust.

Views of Service Users and Carers

The visiting team met three parents who had children with sickle cell anaemia (representing four children in total, ranging in ages between 5 and 24 years), two parents who had children with thalassaemia, and one child with thalassaemia. Overall, the carers were highly complimentary about the care their child received at the hospital, and they were particularly appreciative of the support received from the haemoglobinopathy team. The comments by the user and carers covered the following matters:

- All the parents were highly appreciative of the care they received. They said they felt reassured when both the CNS for haemoglobin disorders and the CNS for benign haematology were on duty, as they knew them well and were confident that any issues they had would be addressed quickly.

- The users and carers considered that their experience of the lead team was that 'nothing was too much trouble' and that they provided care for the whole family, and all the parents considered that they were well informed about their children's health.
- All staff on the day unit and wards were extremely pleasant and always responsive.
- The parents of children with thalassaemia also commended their consultant for 'popping in' to see their children during their transfusions. They felt their consultant was very engaged in their children's treatment.
- The child with thalassaemia enjoyed attending the day unit because there were lots of things to do, like playing games, and said the attending nurses were 'great'.
- Parents of children with sickle cell said their children would benefit from having an individualised care plan that explained their children's needs and medication.
- One carer highlighted issues with their child's allergies not being flagged on the IT system when they had attended for another emergency attendance at the hospital.
- The carers valued the follow-up telephone calls from staff.
- From discussions with the parents, it appeared that the processes for obtaining medication were variable; the parents were unsure why this was the case. Prescriptions for patients with sickle cell disease were sent electronically to the pharmacy department and were often not ready for collection, and parents commented that they often had to return at another time or see their GP. Patients with thalassaemia received their prescriptions on the day unit and did not experience any delays in collecting their medication.
- Children and parents considered that they had a good understanding of the need and also process for having Trans-Cranial Doppler scans (TCD).
- Comments were made that in the few months prior to the visit there had been issues in 'cross matching', with blood samples having to be repeated because of illegible labels on the sample bottles.
- Some carers were not aware of any support from social services (welfare and benefits) or patient involvement opportunities. Others commented that they would find out about services in the local community through other means.
- The parents commented that they felt that the lead consultant and CNSs were very overworked.
- They concluded by saying the entire team worked extremely well together, with one parent saying, "I cannot fault the service my child receives, I think my child's care is one of the best in the UK".

Good Practice

1. Clinical trial activity was impressive considering the shortage of staff within the team. The service was taking part in the STAND clinical trial, looking at the use of crizanlizumab in adolescent and adult sickle cell disease patients.

2. The acute pain team was available on Saturdays to undertake a ward round and review any child who had been admitted with acute pain.

3. Reviewers were impressed with the leadership from the lead clinician and the amount of progress that had been made to improve the quality of the paediatric haemoglobinopathy service since the consultant had commenced in post in 2018.

4. The CNS was actively involved in organising and attending meetings of the 'Northern Nurses' group. The Group consisted of haemoglobinopathy nursing teams from across the northern region and met annually to provide a forum for education and sharing of good practice.

5. The education provision by the hospital school for children who were in-patients was very impressive and had received 'excellent' ratings from OFSTED. Reviewers were particularly impressed that staff would support children who were receiving treatment as in-patients to study for exams and, if necessary, arrange for them to take examinations whilst an in-patient.

Immediate Risks: None were identified during the course of visit.

Concerns

1. **Consultant staffing**

   Reviewers were concerned whether the service had sufficient consultant medical staff with time available for the care of people with haemoglobin disorders to provide staffing for regular reviews, emergency care and clinics. At the time of the visit, the lead consultant had only two programmed activity sessions (PAs) for work with the service. In October 2019 the service had been designated as a Haemoglobinopathy Coordinating Centre (HCC) for Sickle Cell Disease for the north east and Yorkshire, which would also require an increase in the time available for leadership and support to other paediatric haemoglobinopathy services across the region.

   Cover for the lead consultant was from two consultant paediatric haematologists. These three individuals (2.8 wte) were working a 1:3 on call rota to provide 24/7 cover for the benign haematology service. Reviewers were told that a further paediatric haematologist had been recruited who specialised in bone marrow transplantation, and that this could allow some additional time to be allocated to the lead clinician for work with the haemoglobinopathy service.

2. **Clinical Nurse Specialist**

   The haemoglobinopathy CNS had insufficient time (0.4 wte) available for the number of patients being cared for with haemoglobin disorders (approximately 70) to perform clinical, training, leadership and administration tasks for the specialist haemoglobinopathy service. Cover was available from the CNS for benign haematology (1 wte) on the days of the week when the haemoglobinopathy CNS was not working. At the time of the visit the Trust was in the process of recruiting an additional CNS (0.6 wte) for the haemoglobinopathy service.

3. **Access to psychology**

   Psychology staff with appropriate competences in the care of people with haemoglobin disorders were not available. Some support for patients and families was available from the haematology/oncology psychologist on an ‘ad hoc’ basis and also via the general psychology service.

4. **Community support**

   A community service to provide support for patients and their carers within the Sheffield area was not commissioned. The CNSs from the acute Trust team provided some limited support, but were they were unable to provide sufficient time in the community to support children and their families with school care plans, or to support those with more complex needs.

5. **Ward staff competences**

   Evidence was not available to assure the reviewers that staff working with children and young people had competences in caring for children with haemoglobin disorders. A training programme for ward staff had been developed, but the capacity available from the CNSs to provide training for ward staff was limited. Ward nurses did have competences in transfusion skills, and new staff received some formal teaching as part of their induction programme.

6. **Trans-Cranial Doppler Ultrasound Standard Operating Procedure**

   The TCD operating procedure seen at the time of the visit was in a draft format and did not appear to have been reviewed since 2008.

Further Consideration

1. Reviewers were encouraged that there was willingness from the Trust to support the team to develop the SHT and HCC roles, but highlighted the importance of building close links with specialised commissioners in order that the necessary assistance is provided to the team to develop these regional network structures.
2. The CNS team had limited capacity to provide training and education, and were in the process of planning a programme of training with the Trust practice educator. Reviewers considered that identifying 'link' nurses in both the acute and community settings may help with building a wider network of expertise within the Trust and externally.
3. The reviewing team was informed that the laboratory was using 'in-house' reagents to undertake tests such as the sickle solubility test. The review team felt that it would be advisable to consider the use of standardised reagents in such tests in accordance with national policy.
4. Work was being undertaken to upgrade the wards, which, when completed, meant that the wards would only have side rooms and no open bays. Reviewers considered that, as children and young people were used to being cared for in open, more sociable areas such as bays, it may be helpful to do some preparatory work before the move, as some children may have concerns about being in a room on their own.

Commissioning

The team and regional specialist commissioner met for the first time at the peer review visit and were arranging further meetings. However, it is important to note that this report contains a number of issues that would need to be addressed jointly by the Trust and commissioners in order for the paediatric team to function as an effective Specialist Haemoglobinopathy Team (SHT) and fulfil its role as an HCC.

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APPENDIX 1 Membership of Visiting Team

Clinical Lead		
Dr Subarna Chakravorty	Consultant Paediatric Haematologist	King's College Hospital NHS Foundation Trust

Visiting Team		
Amanda Cope	Advanced Nurse Practitioner	Birmingham Women's and Children's NHS Foundation Trust
Dr Jonathan Lancashire	Consultant Paediatric Haematologist	Birmingham Women's and Children's NHS Foundation Trust
Roanna Maharaj	Patient representative	UK Thalassemia Society
Dr Sukhjinder Singh Marwah	Clinical Scientist/ Senior Medical Scientist	The Royal Wolverhampton NHS Trust
Dede-Kossi Osakonor	Highly Specialist Psychologist	Homerton University Hospital NHS Foundation Trust
Giselle Padmore-Payne	Senior Clinical Nurse Specialist for Haemoglobinopathies	Kings College Hospital NHS Foundation Trust
Vanessa Wills	Patient representative	

Quality Review Service		
Sarah Broomhead	Assistant Director	Quality Review Service
Amy Harrison	GMTS	Quality Review Service

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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 – Sheffield Children's NHS Foundation Trust	49	37	76
Total	49	37	76

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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	Service information could be clearer about times of the phlebotomy service and visiting times. 'e' was not applicable as there was no community service in place.
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	

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	However, users and carers who met with the visiting team did not appear to know about any plans for management when attending the ED.
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	N	Information to primary health care team did not include the need for regular prescriptions ('a'), side effects of medication ('b'), guidance for GPs on hydroxycarbamide and iron chelation therapy (if being prescribed by GPs), contraception or sexual health. The clinic letters seen at the time of the visit were not very detailed about the plan of care. The letters did include the patient's medical history.
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	Information would benefit from review to be more explicit about how results would be communicated.

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	Parents who met with the visiting team were not aware of any school care plans and commented that having a school care plan would be very helpful. From the evidence seen and discussion with parents this QS was met for children with thalassaemia.
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 about the transfer of care, including arrangements for monitoring during the time immediately after transition, was not yet in place. Users and carers who met with the visiting team were not clear about the preparation period prior to transfer to the adult service.
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	

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 only had two PAs allocated to the haemoglobinopathy service (clinical and administration). There was no time allocated for leadership of the service or for haemoglobinopathy-related CPD.
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.	N	The lead nurse had only 0.4 WTE time for the haemoglobinopathy service (clinical, administration, training and leadership). Cover for absences was provided by the CNS for inherited and acquired bleeding disorders.
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.	Y	

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	Limited community support was in place. Some support was available from the nurse counsellor. The lead CNS (0.4 wte) had limited time to support children and their families in the community. Evidence that staff had competences in the care of people with haemoglobin disorders was not available. Because of the lack of CNS time, education sessions for staff had not been delivered since 2018. The sessions only lasted between 30 and 60 minutes, which reviewers considered would be insufficient to provide staff with sufficient knowledge about haemoglobin disorders.
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	No specific psychology support for people with haemoglobin disorders was available. Some support was available from the general psychology service and the haematology/oncology psychologist. (British Psychology Society Special Interest Group for Psychologists working in Sickle cell and Thalassaemia (2017) suggests 1 WTE for 300 patients). Access to neuro-psychology was in place.
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 to ensure that all staff were developing and maintaining appropriate competences for their roles in the care of patients with haemoglobin disorders was not yet in place.Training presentations were seen by the reviewers.

Ref	Standard	Met?	Comments
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.	Y	The TCD service was provided by a sonographer who had completed 45 scans.
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	
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 plan was in place to provide training for ED 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	Specialist staff who had expertise in the care of children and young people with haemoglobin disorders were named in the service policy, and there was direct access to all specialties on site.
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	
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	

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 policy could be clearer about the recommended number of cannulation attempts.
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.	Y	
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	

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	Y	'b' not was applicable.
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	
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	All staff had access to the guidelines on the Trust intranet. Staff would contact the haematology team for further advice on management if any haemoglobinopathy patient was admitted.

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 document provided did cover all aspects as defined in the QS, but it was unclear how this linked into wider governance arrangements regarding policy development and document control; for example, there was no authorship, development date or review 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	
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/A	

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/A	All staff who provided support in the community were employed by the Trust.
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 TCD standard operating procedure was out of date. See main report.
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 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.	N	No meeting with representatives of the neonatal screening programme had taken place since 2017.
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	

Ref	Standard	Met?	Comments
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	N	Data were not available covering day unit admissions, ED attendances and readmission rates. The 'did not attend' rate was 19%.
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	

Ref	Standard	Met?	Comments
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	
HN-706	**Network Audits** The service should participate in agreed network-wide audits.	Y	
HN-707	**Research** The SHC should actively participate in research relating to the care of patients with haemoglobin disorders.	Y	The SHT was participating in the crizanlizumab research trial.
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)	Y	Internal quality assurance was undertaken by the lead vascular interventional radiologist, who would review approximately 20% of scans. 'c' was not applicable as a national quality assurance scheme had not yet been established.
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	

Ref	Standard	Met?	Comments
HN-799	**Document Control** All policies, procedures and guidelines should comply with Trust (or equivalent) document control procedures.	Y	

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