# Health Services for People with Haemoglobin Disorders

## Cardiff and Vale University Health Board

Visit Date: 1st October 2019

Report Date: January 2020

8831

## Contents

# Introduction

This report presents the findings of the peer review of health services for people with haemoglobin disorders in Cardiff and Vale University Health Board that took place on 1st 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 (UKFHD) working with the Quality Review Service (QRS). The peer review visit was organised by QRS on behalf of the UKFHD. 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 Health Board'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 Cardiff and Vale University Health Board. 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:

- Cardiff and Vale University Health Board
- Welsh Health Specialist Services Committee

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 organisation 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 organisation in relation to this report is the Cardiff and Vale University Health Board.

## 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 Cardiff and Vale University Health Board 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

## Across the Health Board

**General Comments**

This review looked at the care of adults, children and young people with haemoglobin disorders. During the course of the visit, reviewers met with patients, parents and carers and with staff providing the services, and visited the Emergency Department (ED), day unit and wards.

At the time of the visit the Cardiff and Vale University Health Board provided health services for over 500,000 people living in Cardiff and the Vale of Glamorgan.

The University Hospital of Wales and The Noah's Ark Children's Hospital were reviewed as Specialist Haemoglobinopathy Teams (SHTs) for adults and paediatric services respectively. Both teams provided specialist care for patients residing within five health boards across South and Mid Wales (see table 1 below).

Around three to four babies were born in Wales each year with sickle cell disease (SCD), and one baby every two years with thalassaemia. Nearly all children born in Wales affected with SCD, and the majority of those affected with thalassaemia, were identified by neonatal testing or newborn bloodspot screening programmes.

Other new patients presented through immigration or late diagnosis. There was a slight annual fluctuation of adult patient numbers depending on the student population at university, as the team provided a haemoglobinopathy service for the whole of South Wales.

In 2014 the community sickle cell and thalassaemia services moved to a multi-purpose community hub based at Loudoun Square.

Some of the reviewer findings for action across the Health Board were the same for both the adult and the paediatric services and have therefore been repeated in each section.

**Table 1**

| Specialist Haemoglobinopathy Centre | Linked Health Boards and hospitals |
|---|---|
| Cardiff and Vale University Health Board<br>• University Hospital of Wales<br>• The Noah's Ark Children's Hospital for Wales | Cardiff & Vale University Health Board<br>• University Hospital Llandough (no A+E)<br>• Cardiff Royal Infirmary<br>• Barry Hospital<br>• Whitchurch Hospital |
| | Swansea Bay University Health Board<br>• Morriston Hospital, Swansea<br>• Princess of Wales Hospital, Bridgend |
| | Aneurin Bevan University Health Board<br>• Nevill Hall Hospital, Abergavenny<br>• Royal Gwent Hospital, Newport |
| | Cwm Taf University Health Board<br>• Prince Charles Hospital, Merthyr Tydfil<br>• Royal Glamorgan Hospital, Llantrisant |
| | Powys Teaching Health Board<br>No major A+E departments |
| | Hywel Dda University Health Board<br>• Bronglais General Hospital, Aberystwyth |

| Specialist Haemoglobinopathy Centre | Linked Health Boards and hospitals |
|---|---|
| | • Glangwili General Hospital, Carmarthen<br>• Withybush General Hospital, Haverfordwest |

**ADULTS**

| Health Board | Reviewed as: | No. of adults with sickle cell disease | No. of adults with thalassaemia | No. of adults on long-term red cell transfusions |
|---|---|---|---|---|
| Cardiff and Vale University Health Board - University Hospital of Wales | SHT | 46 | 14 | 14 |

**CHILDREN AND YOUNG PEOPLE**

| Health Board | Reviewed as: | No. of children with sickle cell disease | No. of children with thalassaemia | No. of children on long-term red cell transfusions |
|---|---|---|---|---|
| Cardiff and Vale University Health Board - The Noah's Ark Children's Hospital for Wales | SHT | 38 | 15 | 3 |

| Support groups available in the Cardiff area for patients and carers | Y/N |
|---|---|
| Sickle Cell Disease – Adults | Y |
| Thalassemia – Adults | Y |
| Sickle Cell Disease – Children | Y |
| Thalassemia – Children | Y |

**Progress since last visit**

The previous review had taken place in 2016, and since this time there had been significant progress and a number of improvements:-

- Appointment, as clinical lead of the adult service, of a consultant haematologist based at the hospital
- Introduction of clear pathways for adults in the management of painful crisis and other presentations, with streaming of acute patients through 'majors' area in the ED
- Introduction of emergency and elective automated red cell exchange for both adults and children
- Regular teaching sessions for ED and haematology staff, including an annual MDT education and awareness day
- Introduction of a transition clinic
- Joint Laboratory/Clinical/Medical Genetics MDT commenced in 2015
- Adult clinical guidelines for patients with sickle cell disease reviewed
- HESTIA 3 & 4 trials for children opened in 2018
- Part time support worker for children with haemoglobin disorders appointed in conjunction with the Ethnic Minorities & Youth Support Team (EYST), funded through a BBC Children in Need grant

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## Specialist Team (Adult Services): Cardiff and Vale University Health Board

**General Comments and Achievements**

Patients who met with the visiting team were appreciative of the care they received from the haemoglobinopathy team, which was led by a consultant haematologist and a clinical nurse specialist based at the University Hospital of Wales. Having an on-site named lead was a welcome improvement in the adult service since the last peer review visit, as there had only been limited support from a visiting consultant at that time. The lead team had a good vision for the future development of the service, and was exploring more formalised links with some English Specialist Haemoglobinopathy Teams (SHTs). The team was proud of what they had achieved since the last visit in 2016, and it was clear to the reviewers that they were highly committed and enthusiastic.

The clinical nurse specialist (CNS) provided specialist support for patients with non-malignant haematology conditions including inherited anaemias, and was based mainly at the University Hospital of Wales and in the Sickle Cell Centre at Loudoun Square, providing advice and input to the out-patient service.

Patients requiring regular transfusions were reviewed and treated in the haematology day centre, which also provided all the out-patient chemotherapy and supportive care for patients with haematological malignancies. There was no facility to provide routine out-of-hours transfusions.

Both the adult haematology consultant and the CNS had engaged with national haemoglobinopathy networks for education: for example, the UKFHD had hosted a meeting in Cardiff, and work had been done with the Sickle Cell and UK Thalassaemia Societies. The CNS was also making links with CNSs from other SHTs.

**Staffing**

| Staffing for an SHT Adult Haemoglobinopathy Service [1] | Number of patients | Actual wte (at time of the visit) | Staffing required as recommended by NHSE |
|---|---|---|---|
| Consultant haematologist with >0.6 WTE per 150 patients dedicated to work with patients with haemoglobinopathies | 60 | 0.1 | 0.24 |
| At least 0.25 WTE allocated to haemoglobinopathies CPD in the adult consultant job plan | 60 | 0.1 | - |
| Clinical psychologist for adult patients with >0.5 WTE per 200 patients dedicated to work with patients with haemoglobinopathies[2] | 60 | 0 | 0.15 |

**Emergency Care**

Patients known to the service were given information with contact numbers for the CNS, day centre and haematology ward. Patients needing emergency care could contact the CNS during normal working hours and the ward at other times. The hospital aimed to admit patients directly to the ward where possible, but in-patient beds were not always available and so patients were sometimes advised to attend the ED.

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

[2] *The Quality Standard for psychology staffing (HN-205) references the recommendation from the British Psychology Society Special Interest Group for Psychologists working in Sickle Cell and Thalassaemia (2017) which suggests 1 wte for every 300 patients.*

**In-patient Care**

The adult in-patient ward was a 27-bedded dedicated haematology/oncology unit with ten beds designated for bone marrow transplants.

**Day care**

The adult haematology day centre provided out-patient treatment for patients with haematological malignant and non-malignant conditions. The day centre provided red cell transfusions and red cell exchanges for patients with haemoglobinopathies. At the time of the visit the day centre was housed in temporary accommodation because of maintenance work on the main unit.

**Community-based Care**

Patients were able to access a weekly drop-in session at The Sickle Cell and Thalassaemia Centre, based at the Loudoun Square facility in Butetown.

**Views of Service Users and Carers**

The visiting team met with eight users and carers during the course of the visit, and drew the following conclusions from these meetings:

- Overall, the users and carers were highly complimentary about the care and support available from the lead clinician and CNS, and considered that the service had improved.
- The users and carers considered that the CNS was 'amazing': she was always available for advice and support and 'held the service together'. Some patients commented that she was often available for advice outside normal working hours, and that they were worried when they knew she was going to be away.
- None of the patients who met with the reviewers had received any written information about their condition or been given a copy of their care plan or summary of their annual review.
- All those who met with the reviewers considered that the lack of access to psychology support was an issue, and that they would benefit from better access to these services.
- Patients commented that they were not aware of any community services from which they could receive additional support and advice, though they could attend a drop-in session run by the CNS.
- Users and carers were not confident that staff in the ED had sufficient understanding of haemoglobin disorders, commenting that patients often had to wait for appropriate analgesia.
- Patients with thalassaemia who met with the reviewing team were asked about the monitoring of their condition and the need for regular diagnostic measures such as DEXA and MRI scans, but did not think that these had been performed. They were also not clear about whether regular endocrinology blood tests were being done as they did not receive any written information or feedback.

**Good Practice**

1. Reviewers were impressed with the implementation of an integrated pathway through the ED. Patients attending the ED would be seen in the 'majors' area initially, and quickly referred to the on-call haematologist for ongoing care. The integrated pathway had significantly improved the time to initial assessment, treatment by a senior decision maker and then handover to the clinical team. All known patients with haemoglobin disorders had an emergency care plan documented which could be accessed by ED staff.

2. It was evident to reviewers who visited the ED during the visit that there was good leadership and engagement from the ED lead and the team working within the department, in respect of improving the pathway for patients with haemoglobin disorders. Reviewers who had been part of the review team in 2016 commented that this was an impressive change from their experience three years ago.

3. The hospital team had a good working relationship with the senior management team and the Welsh Health Specialist Services Committee. Reviewers met with representatives from these groups, and these discussions showed that the proposed strategic plan to develop the SHT was encouraging.

**Immediate Risks:** None were identified at the time of the visit.

**Concerns**

1. **Patient information and access to care plans**

   Patients did not receive any written information, copies of their care plan or clinic letters, or a summary of their annual review. Written information about their condition and care treatment decisions should be available to ensure that patients have sufficient knowledge to give informed consent. Reviewers were concerned that this issue had been identified as a concern during the last visit in 2016 and had not yet been addressed.

2. **Consultant staffing**

   Reviewers were concerned that the lead consultant had insufficient time available for the care of people with haemoglobin disorders to provide leadership and governance for the service as well staffing for regular reviews, emergency care and clinics. At the time of the visit the consultant had only one programmed activity (PA) session for work with the service, and there was no cover for absence. This lack of consultant time meant that it was very difficult for further service development to occur to enable the national standards to be met.

3. **Clinical nurse specialist cover**

   The CNS provided specialist support for all adult patients with non-malignant haematological conditions, both in the hospital and in the community, and had no cover for absence. Patients who met with the visiting team were concerned about the vulnerability of the service, as they considered the CNS to be pivotal to the smooth running of the service.

4. **Access to psychology**

   Access to psychology staff with appropriate competences in the care of people with haemoglobin disorders and neuropsychology was not available.

5. **Clinical guidelines – thalassaemia**

   Guidelines covering the management of patients with thalassaemia were not yet in place.

6. **Clinical guidelines – sickle cell disease**

   Many of the guidelines had been updated before the visit but their detail was limited. The guidance covering indications for drug therapy for patients on hydroxycarbamide were not consistent with the British Society for Haematology Guidance (2018). The guidance on blood transfusions was also inconsistent with the Health Board guidelines. The guidance did not include contact details for specialist and support services, though the lead consultant haematologist's and CNS's contact details were usually listed for advice and guidance. Reviewers considered that the governance process for the development of clinical guidelines would benefit from being strengthened to ensure that guidance is appropriately governed and referenced.

7. **Multidisciplinary discussion**

   Multidisciplinary team (MDT) meetings to discuss and review patient care were no longer taking place. In-patients were discussed as part of the general haematology/oncology MDT, but there was no formal mechanism for the multidisciplinary discussion of complex and community-based patients. Reviewers considered that it would be important to re-establish these meetings as soon as possible.

**Further Consideration**

1. A number of the issues raised in this report were symptomatic of a lead team who were working very hard but had insufficient time and capacity allocated to provide a comprehensive specialist haemoglobinopathy service (development of guidelines and patient information, staff and patient education and audit). Reviewers were very clear that since the last visit significant progress had been made in some areas but not in others. Reviewers were concerned that, without appropriate support from senior management and commissioners, the situation was unlikely to improve.
2. The clinical team recognised the need to establish collaborative links with other specialist centres with higher patient numbers in order to enhance their expertise, and the review team supported this endeavour. Reviewers considered that benefits in formalising networking arrangements would enable:-
    a. a wider haemoglobinopathy MDT to discuss and review patient care, especially for those with more complex needs;
    b. improved pathways to specialist services by formalising links to named specialists; and
    c. sharing of information and improved collaborative working especially in terms of review and learning, developing patient information, clinical guidance and competence frameworks for staff.
3. A service organisation policy was not yet in place. Development of a service organisation policy would help define a roadmap for day-to-day operation, give guidance for decision-making, and streamline internal processes.
4. Patients had no access to an out-of-hours service for routine blood transfusions.
5. An audit programme was not yet in place, including audit of time to analgesia.
6. In the light of the comments received by the patients with thalassaemia at the time of the visit, it may be helpful to review the process for communicating and providing information for patients to ensure that they have sufficient knowledge about their condition.
7. The transition process was not yet fully implemented to ensure that all young people would be given the appropriate information and support to transition to adult care. Transition clinics were held on an ad hoc basis, and the adult and paediatric teams had plans to implement the nationally-adopted ‘Ready Steady Go’ transition programme.
8. NHS Wales had signed an agreement to enable patients with haemoglobin disorders to be registered on the National Haemoglobinopathy Register (NHR), but reviewers were told that registering patients on the NHR was hampered by the lack of time available from the clinical team and the lack of data management support.

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## Specialist Team (Children and Young People Services): Cardiff and Vale University Health Board

**General Comments and Achievements**

The paediatric haemoglobinopathy service was part of the Paediatric Haematology/Oncology department. The Paediatric Haematology/Oncology unit at the Children's Hospital for Wales served the entire population of children under the age of 16 in South, Mid and West Wales. At the time of the visit there were no other hospitals or facilities in Wales that provided a haemoglobin disorder service. Patients in North Wales were transferred to England to be treated in Alder Hey Children's Hospital, Liverpool (see table 1 on page 5).

At the time of the visit, the service was led by a consultant paediatric haematologist with support from a deputy lead consultant paediatric haematologist and with additional cover available from the consultant paediatric oncologists. The CNS had been on long-term leave, and in the interim there had been some cover from other members of the paediatric nursing team. It was clear to the reviewers that, whilst the team were enthusiastic and proactive in improving patient care, they were also working within staffing constraints.

Transition clinics had commenced, with young people being offered a tour of the adult services. The apheresis programmes had been expanded and children and young people were being referred for stem cell transplantation.

Children and parents who met with the visiting team were appreciative of the care they received.

**Staffing**

| Staffing for an SHT Paediatric Haemoglobinopathy Service[3] | 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 | 43 | 0.1 | 0.17 |
| At least 0.25 wte allocated to haemoglobinopathies CPD in the paediatric consultant job | 43 | 0 | - |
| Clinical psychologist for paediatric patients with >0.5 wte per 150 patients dedicated to work with patients with haemoglobinopathies | 43 | 0 | 0.14 |

**Emergency Care**

Patients requiring an acute assessment had open access to Rainbow Ward. Children who presented at the Emergency Department (ED) were seen by the on-call paediatrician.

**Inpatient Care**

Children with haemoglobinopathies were admitted to Rainbow Ward, a 13-bedded oncology ward. The Teenage Cancer Trust Unit (TCTU) could also admit young people with haemoglobin disorders if there was capacity.

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

**Day care**

Rainbow Day Unit had five beds and two treatment chairs (including an anaesthetic room with one general anaesthetic list for day procedures each week). The unit was open between the hours of 9am and 6pm, from Monday to Friday. Simple top-up transfusions were managed on the day unit. Patients who required exchange transfusions were admitted to Rainbow Ward.

**Out-patient care**

Non-malignant haematology clinics were held in Rocket Ward, which was dedicated to haematology/oncology children's out-patients on Monday mornings. Trans-Cranial Doppler (TCD) clinics were undertaken by a vascular scientist from the University Hospitals Bristol NHS Foundation Trust. Because of the low numbers of young people requiring transition to adult services the joint transition clinics were arranged on an ad hoc basis.

**Community-based Care**

At the time of the review, the paediatric services were not supported by any community-based care.

**Views of Service Users and Carers**

During the course of the visit, the visiting team only met three children and young people with thalassaemia who were receiving treatment, together with their parents. The team had invited some children and families with sickle cell disease to meet with the reviewing team, but unfortunately these families were unable to attend. Overall, the children, young people and parents were appreciative and happy about the treatment they received whilst attending for blood transfusions. The children, young people and families who spoke to the reviewing team expressed concerns over not being given enough information about the patient's condition. However, all present spoke highly of their medical and nursing teams and loved being on the day unit. It was also good to note that interpreters were available when required.

**Good Practice**

1. Reviewers were impressed that a physiotherapist attended the annual review and TCD clinics. Children and young people had a musculoskeletal assessment and a respiratory function test so that advice could be given and effective early interventions, if required, could be commenced, to help reduce the risk of further complications at a later age.
2. The teenage room was well designed with a recreational area. Young people with haemoglobin disorders who required in-patient care could be admitted to the unit if capacity allowed.
3. The service was actively engaged in the HESTIA 3 & 4 trials for children, which opened in 2018.
4. The EYST Saturday club for children and young people affected by sickle cell disease and thalassaemia provided lots of advice, education, social support and activities for children and young people, as well as support to their families.

**Immediate Risk:** None were identified at the time of the visit

**Concerns**

1. **Patient information and access to care plans**

   Patients and parents did not receive any written information (including information on TCD), copies of their care plans, clinic letters, or summaries of their annual reviews. There was no evidence of any paediatric patient information for haemoglobinopathies. Written information about patients' conditions and care treatment decisions should be available to ensure that they have sufficient knowledge to give informed consent. Reviewers were concerned that this issue had been identified during the last visit in 2016 and had not yet been addressed.

2. **Consultant staffing**

   Reviewers were concerned that the lead consultant had insufficient time for available for the care of people with haemoglobin disorders to provide leadership and governance for the service as well staffing for regular reviews, emergency care (1:2 on-call rota) and clinics. At the time of the visit, the lead consultant and deputy had only 0.5 programmed activity (PA) each for their work with the service.

3. **Clinical Nurse Specialist support**

   At the time of the visit the CNS was on long-term leave, and there was limited cover available from the paediatric oncology CNS team. A paediatric community service for children and young people with haemoglobin disorders was not in place and, in the absence of the CNS, no health input was available into the development of school care plans.

4. **Access to psychology**

   Access to psychology staff with appropriate competences in the care of people with haemoglobin disorders and neuropsychology was not available.

5. **Clinical guidelines – sickle cell disease**

   The guidelines covering acute care of children and young people with sickle cell disease had been updated shortly before the review and were not yet available on the Health Board intranet. The draft guidelines did not include the latest national best practice guidance; for example, the clinical guidance regarding pre-operative transfusion in sickle cell disease did not include the current recommendation of transfusing patients to a target haemoglobin concentration of 100g/l. The guidance also included the taking of arterial blood gas as part of the monitoring of children who are acutely unwell, which is not deemed best practice.

6. **Clinical guidelines – thalassaemia**

   Guidelines covering the care of children and young people with thalassaemia were not yet in place.

7. **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. Ward nurses did have competences in transfusion skills, and new staff did receive some formal teaching as part of their induction programme.

8. **Multidisciplinary discussion**

   There was no formal mechanism for multidisciplinary discussion of in-patients or complex and community-based patients. Reviewers considered that it would be important to establish these meetings as soon as possible.

**Further Consideration**

1. A number of the issues raised in this report were symptomatic of a lead team who were working very hard but had insufficient time and capacity allocated to provide a comprehensive specialist haemoglobinopathy service (development of guidelines and patient information, staff and patient education and audit). Reviewers were very clear that since the last visit significant progress had been made in some areas but not in others. Reviewers were concerned that, without appropriate support from senior management and commissioners, the situation was unlikely to improve.
2. The clinical team recognised the need to establish collaborative links with other paediatric specialist teams in order to enhance their expertise, and the review team supported this endeavour. Reviewers considered that benefits in formalising networking arrangements would enable:-
   a. a wider haemoglobinopathy MDT to discuss and review patient care, especially for those with more complex needs;

b. improved pathways to specialist services by formalising links to named referrers; and

c. sharing of information and improved collaborative working, especially in terms of review and learning, developing patient information, clinical guidance and competence frameworks for staff.

3. A service organisation policy was not yet in place. Development of a service organisation policy would help define a roadmap for day-to-day operation, give guidance for decision-making, and streamline internal processes.

4. Patients had no access to an out-of-hours service for routine blood transfusions.

5. The service had not completed all the recommended audits. Audits covering 'time to analgesia' and the numbers of admissions for the sickle cell screening programme had been completed.

6. The transition process was not yet fully implemented to ensure that all young people would be given the appropriate information and support to transition to adult care. Transition clinics were held on an ad hoc basis, and the adult and paediatric teams had plans to implement the nationally-adopted 'Ready Steady Go' transition programme.

7. Data submitted in advance of the visit suggested that all patients had had an annual review in the last year, but in the five clinical records seen during the visit there was no evidence filed that an annual review had taken place.

8. The TCD service was provided by an external provider. Although quality assurance for the service is the responsibility of the provider, it may be helpful to review the service level agreement to check that it covers all the requirements of the Quality Standard (HN-606).

9. NHS Wales had signed an agreement to enable patients with haemoglobin disorders to be registered on the National Haemoglobinopathy Register (NHR), but reviewers were told that registering patients on the NHR was hampered by the lack of time available from the clinical team and the lack of data management support.

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## Commissioning

The services had good links with specialised commissioners, and discussions with the reviewers clearly showed there was a willingness between providers and commissioners to develop both the adult and the paediatric haemoglobinopathy services.

This report, however, contains a number of issues, some of which were highlighted in the previous report (2016). These will need to be addressed, with commissioner support, in order for both the adult and the paediatric teams to function as comprehensive Specialist Haemoglobinopathy Teams (SHT). In the meeting with the commissioners it was suggested that forming a link with a Haemoglobinopathy Coordinating Centre to provide support, and increasing the time dedicated to haemoglobinopathies in the senior team's job descriptions, would be essential to enable Cardiff to progress to become a comprehensive specialist centre for these disorders in Wales.

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# APPENDIX 1 Membership of Visiting Team

| Clinical Leads | | |
|---|---|---|
| Dr Subarna Chakravorty | Consultant Paediatric Haematologist | King's College Hospital NHS Foundation Trust |
| Dr Emma Drasar | Consultant Haematologist | Whittington Health NHS Trust |

| Visiting Team | | |
|---|---|---|
| Halima Begum | Quality Manager for Clinical Haematology | Imperial College Healthcare NHS Trust |
| Sandy Hayes | Adult Haemoglobinopathy Senior Specialist Nurse and Thames Valley Network Lead Nurse | Oxford University Hospitals NHS Foundation Trust |
| Janice Llewellyn | Paediatric Haematology/Oncology Nurse Specialist | The Shrewsbury & Telford Hospital NHS Trust |
| Roanna Maharaj | Patient Representative | UK Thalassemia Society |
| Rachel McFee | Chief Executive OSCAR Patients Group | OSCAR Sandwell |
| Professor Corrina McMahon | Consultant Haematologist | Our Lady's Children's Hospital, Crumlin, Dublin |
| Dr Elizabeth Rhodes | Consultant Haematologist | St George's University Hospitals NHS Foundation Trust |
| Maureen Scarlett | Community Nurse Specialist Haemoglobinopathies | Cambridgeshire Community Services NHS Trust |
| Lesley McCarthy | Roald Dahl Haemoglobinopathy Nurse Specialist | Oxford University Hospitals NHS Foundation Trust |

| QRS Team | | |
|---|---|---|
| Sarah Broomhead | Assistant Director | 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 |
|---|---|---|---|
| Adults | 38 | 14 | 37 |
| Paediatrics | 45 | 14 | 31 |
| **Total** | **83** | **28** | **34** |

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## Specialist Services for People with Haemoglobin Disorders

| Ref | Standard | Adults | | Paediatrics | |
|---|---|---|---|---|---|
| | | Met? | | Met? | Comments |
| HN-101 | **Haemoglobin Disorder Service Information**<br><br>Written information should be offered to patients and, where appropriate, their carers covering at least:<br>a. Brief description of the service, including times of phlebotomy, transfusion and psychological support services<br>b. Clinic times and how to change an appointment<br>c. Ward usually admitted to and its visiting times<br>d. Staff of the service<br>e. Community services and their contact numbers<br>f. Relevant national organisations and local support groups<br>g. Where to go in an emergency<br>h. How to:<br>i. Contact the service for help and advice, including out of hours<br>ii. Access social services<br>iii. Access benefits and immigration advice<br>iv. Interpreter and advocacy services, PALS, spiritual support and HealthWatch (or equivalent)<br>v. Give feedback on the service, including how to make a complaint<br>vi. Get involved in improving services (QS HN-199) | N | Written information was not yet in place covering the requirements of the QS. Patients who met with the visiting team confirmed that they did not receive any written information, but did receive verbal information about services from members of the team. The CNS had a contact card that was given to patients. | N | Written information was not yet in place covering the requirements of the QS. |

| Ref | Standard | Adults | | Paediatrics | |
|---|---|---|---|---|---|
| | | Met? | | Met? | Comments |
| HN-102 | **Information about Haemoglobin Disorders**<br><br>Written information should be offered to patients and, where appropriate, their carers covering at least:<br>a. A description of their condition (SC or T), how it might affect them and treatment available<br>b. Problems, symptoms and signs for which emergency advice should be sought<br>c. How to manage pain at home (SC only)<br>d. Splenic palpation and Trans-Cranial Doppler scanning (children only)<br>e. Transfusion and iron chelation<br>f. Possible complications, including priapism and complications during pregnancy<br>g. Health promotion, including:<br>i. Travel advice<br>ii. Vaccination advice<br>h. National Haemoglobinopathy Registry, its purpose and benefits<br>i. Self-administration of medications and infusions | N | As QS HN-101. | N | As QS HN-101. |

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| HN-103 | **Care Plan**<br><br>All patients should be offered:<br>a. An individual care plan or written summary of their annual review including:<br>i. Information about their condition<br>ii. Plan for management in the Emergency Department<br>iii. Planned acute and long-term management of their condition, including medication<br>iv. Named contact for queries and advice<br>b. A permanent record of consultations at which changes to their care are discussed<br>The care plan and details of any changes should be copied to the patients' GP and their local / specialist team consultant (if applicable). | N | Patients were not given a copy of their care plan, clinic letters or a summary of their annual review. From the patient records seen at the visit, GPs did not receive copies of patients' annual reviews. | N | Patients were not given a copy of their care plan, clinic letters or a summary of their annual review. |
| HN-104 | **Information for Primary Health Care Team**<br><br>Written information should be sent to the patient's primary health care team covering available local services and:<br>a. The need for regular prescriptions including penicillin or alternative (SC and splenectomised T) and analgesia (SC)<br>b. Side effects of medication, including chelator agents [SC and T]<br>c. Guidance for GPs on:<br>i. Hydroxycarbamide and iron chelation therapy (if being prescribed by GPs)<br>ii. Immunisations<br>iii. Contraception and sexual health<br>d. Indications and arrangements for seeking advice from the specialist service | N | The information sent to GPs did not cover the side effects of medications ('b'), guidance on contraception and sexual health ('c' iii), or indications and arrangements for seeking advice from the specialist team ('d').<br>'a' and 'c' i were not applicable as GPs did not prescribe therapy, although reviewers considered that GPs should receive some information covering regular medication. Information to GPs did cover immunisations ('c' ii). | N | The information sent to GPs did not cover side effects of medications ('b'), guidance on contraception and sexual health ('c' iii), or indications and arrangements for seeking advice from the specialist team ('d').<br>'a' and 'c' i were not applicable as GPs did not prescribe therapy, although reviewers considered that GPs should receive some information covering regular medication. Information to GPs did cover immunisations ('c' ii). |

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| HN-105 | **Information about Trans-Cranial Doppler Ultrasound (Children's Services Only)**<br><br>Written information should be offered to patients and their carers covering:<br>a. Reason for the scan and information about the procedure<br>b. Details of where and when the scan will take place and how to change an appointment<br>c. Any side effects<br>d. Informing staff if the child is unwell or has been unwell in the last week<br>e. How, when and by whom results will be communicated | N/A | | N | Written information was not yet in place covering the requirements of the QS. |
| HN-106 | **School Care Plan (Children's Services Only)**<br><br>A School Care Plan should be agreed for each child or young person covering, at least:<br>a. School attended<br>b. Medication, including arrangements for giving / supervising medication by school staff<br>c. What to do in an emergency whilst in school<br>d. Arrangements for liaison with the school<br>e. Specific health or education need (if any) | N/A | | N | There was no evidence of any school care plans in the notes seen at the time of the visit. Reviewers were told that the CNS would liaise with schools in supporting children with medical needs. |
| HN-194 | **Environment**<br><br>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 | | Y | |

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| HN-195 | **Transition to Adult Services**<br><br>Young people approaching the time when their care will transfer to adult services should be offered:<br>a. The opportunity to discuss the transfer of care at a joint meeting with paediatric and adult services<br>b. A named coordinator for the transfer of care<br>c. A preparation period prior to transfer<br>d. Written information about the transfer of care including arrangements for monitoring during the time immediately afterwards | N | From the evidence seen and discussion with patients, written information about the transfer of care (including arrangements for monitoring during the time immediately afterwards) was not yet in place.<br>All other aspects of the QS were met. | N | From the evidence seen and discussion with patients, written information about the transfer of care (including arrangements for monitoring during the time immediately afterwards) was not yet in place.<br>All other aspects of the QS were met. |
| HN-199 | **Involving Patients and Carers**<br><br>The service's involvement of patients and carers should include:<br>a. Mechanisms for receiving feedback from patients and carers<br>b. An annual patient survey (or equivalent)<br>c. Mechanisms for involving patients and, where appropriate, their carers in decisions about the organisation of the service<br>d. Examples of changes made as a result of feedback and involvement of patients and carers | Y | | Y | |
| HN-201 | **Lead Consultant**<br><br>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 1 PA allocated to the haemoglobinopathy service (clinical, administration and geographical lead). There was no cover in place for absences. | N | The lead consultant only had 0.5 PA allocated to the haemoglobinopathy service (clinical, administration and geographical lead). |

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| HN-202 | **Lead Nurse**<br><br>A lead nurse should be available with:<br>a. Responsibility, with the lead consultant, for guidelines, protocols, training and audit relating to haemoglobin disorders<br>b. Responsibility for liaison with other services within the network<br>c. Competences in caring for people with haemoglobin disorders<br>d. Competences in the care of children and young people (children's services only)<br>The lead nurse should have appropriate time for their leadership role and cover for absences should be available. | N | The lead nurse did not have cover for absences or time allocated for liaison with other services. All other aspects of the QS were met. | N | Evidence was not available to demonstrate that the lead CNS had competences in caring for children with haemoglobin disorders. The lead CNS did not have time allocated for liaison with other services or for CPD. At the time of the visit the lead CNS was on long term leave. Some cover was available from the oncology CNSs. |
| HN-203 | **Medical Staffing and Competences**<br><br>The service should have sufficient medical staff with appropriate competences in the care of people with haemoglobin disorders, including:<br>**All services:**<br>a. Haematology or paediatric medical staffing for clinics and regular reviews<br>b. 24/7 consultant and junior staffing for emergency care<br>**SHCs only:**<br>c. A consultant specialising in the care of people with haemoglobin disorders on call and available to see patients during normal working hours<br>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<br>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 cover was available out of hours. In practice, the lead consultant provided 2nd on-call cover but was also contacted when away for advice. Cover for the lead clinician was from another haemato-oncologist, and it was not clear that they had competences in caring for adults with haemoglobin disorders. | Y | Cover for the lead clinician was from other paediatric oncologists, and it was not clear that two of these consultants had competences in caring for children and young people with haemoglobin disorders. |

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| HN-204 | **Nurse Staffing and Competences**<br><br>The service should have sufficient nursing staff with appropriate competences in the care of people with haemoglobin disorders, including:<br>a. Clinical nurse specialist/s with responsibility for the acute service<br>b. Clinical nurse specialist/s with responsibility for the community service<br>c. Ward-based nursing staff<br>d. Day unit (or equivalent) nursing staff<br>e. Nurses with competences in cannulation and transfusion available at all times patients attend for transfusion<br>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 was not yet in place, and there was no evidence that ward staff had completed competences in haemoglobin disorders.<br>New staff did receive some formal teaching as part of their induction.<br>'a' and 'b' were met in terms of competences, but the CNS did not have any cover for absences. | N | There was no evidence that staff working with children and young people had competences in caring for children with haemoglobin disorders.<br>Reviewers were told that the ward nurses did have competences in transfusion skills, but the evidence available was not up to date and the ward manager could not provide an up to date list as the information was held centrally by the Trust.<br>New staff did receive some formal teaching as part of their induction programme.<br>Some useful information was available in the resources folder.<br><br>A community service was not yet in place. |

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| HN-205 | **Psychology Staffing and Competences**<br><br>The service should have sufficient psychology staff with appropriate competences in the care of people with haemoglobin disorders, including:<br>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<br>b. Time for input to the service's multi-disciplinary discussions and governance activities<br>c. Provision of, or arrangements for liaison with and referral to, neuro-psychology<br>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. (British Psychology Society Special Interest Group for Psychologists working in Sickle Cell and Thalassaemia (2017) suggests 1WTE for 300 patients). | N | No specific psychology support for people with haemoglobin disorders was available. (British Psychology Society Special Interest Group for Psychologists working in Sickle Cell and Thalassaemia (2017) suggests 1WTE for 300 patients). |
| HN-206 | **Training Plan**<br><br>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. | 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. |
| HN-207 | **Trans-Cranial Doppler Ultrasound Competences (Children's Services Only)**<br><br>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 | | Y | The TCD service was provided on site from a practitioner from University Hospitals Bristol NHS Foundation Trust |

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| HN-299 | **Administrative, Clerical and Data Collection Support**<br><br>Administrative, clerical and data collection support should be appropriate for the number of patients cared for by the service. | Y | | N | No administrative or clerical support was available to help with data management. |
| HN-301 | **Support Services**<br><br>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.<br>a. Social worker/ benefits adviser<br>b. Leg ulcer service<br>c. Play specialist (children's services only)<br>d. Chronic pain team (adult services only)<br>e. Dietetics<br>f. Physiotherapy (in-patient and community-based)<br>g. Occupational therapy<br>h. Mental health services (adult and CAMHS)<br>i. DNA studies<br>j. Polysomnography | N | The service did not have access to a social worker/ benefits adviser. All other aspects of the QS were met. | N | The service did not have access to a social worker/ benefits adviser. There was access to a dietitian, a physiotherapist and an mental health nurse (RMN) for in-patient paediatric patients if required. The ward manager reported that the support from the RMN was very good, although the support from the local CAMHS service was limited.<br>Access to a hospital play specialist was available on both the day care unit and the ward, Monday to Friday.<br>There was also access to polysomnography. |
| HN-302 | **Emergency Department – Staff Competences**<br><br>Medical and nursing staff working in the Emergency Department/s should have competences in urgent care of people with haemoglobin disorders. | Y | | Y | |

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| HN-303 | **Specialist On-site Support**<br><br>Access to the following specialist staff and services should be easily available on the same hospital site as the specialist team:<br>a. Manual exchange transfusion (24/7)<br>b. Erythrocytapheresis<br>c. Acute pain team including specialist monitoring of patients with complex analgesia needs<br>d. High dependency care, including non-invasive ventilation<br>e. Level 2 and 3 critical care | Y | | Y | |

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| HN-304 | **Specialist Services - Network**<br><br>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:<br>a. Pulmonary hypertension team (adults)<br>b. Consultant obstetrician with an interest in care of people with haemoglobin disorders and specialist high risk anaesthetics (adults)<br>c. Respiratory physician with interest in acute/chronic sickle lung disease and obstructive sleep apnoea (adults & children)<br>d. Fertility, including pre-implantation genetic diagnosis and sperm storage (adults)<br>e. Consultant cardiologist with interest in sickle cardiomyopathy, iron overload related heart disease (adults)<br>f. Consultant endocrinologist with interest in thalassaemia related endocrinopathy and osteoporosis (adults)<br>g. Consultant paediatric endocrinologist with interest in growth problems related to haemoglobinopathies and thalassaemia related endocrinopathy (children)<br>h. Hepatobiliary team with an interest in sickle hepatopathy, viral liver disease, iron overload-related liver disease (adults & children)<br>i. Consultant neurologist and neurosurgeon with an interest in sickle vasculopathy (adults & children)<br>j. Hyperacute stroke service (adults)<br>k. Consultant ophthalmologist with an expertise in sickle retinopathy and chelation related eye disease (adults & children)<br>l. Consultant nephrologist with expertise in sickle nephropathy (adults & children)<br>m. Consultant urologist with expertise in managing priapism and erectile dysfunction (adults & children)<br>n. Orthopaedic service with expertise in managing sickle and thalassaemia related bone disease (adults & children)<br>o. Specialist imaging, including<br>i. MRI tissue iron quantification of the heart and liver<br>ii. Trans-Cranial Doppler ultrasonography (children)<br>p. Bone marrow transplantation services (children only)<br>q. Physiotherapy services (in patient and community based)<br>r. Interventional and neuroradiology for neurovascular complications | N | Reviewers were told that specialist services were available from London or Birmingham, but the staff providing these services were not named and it was not clear that indications for referral had been formalised. | N | Reviewers were told that specialist services were available from London or Birmingham, but the staff providing these services were not named and it was not clear that indications for referral had been formalised. |

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| HN-305 | **Laboratory Services**<br><br>UKAS / CPA accredited laboratory services with satisfactory performance in the NEQAS haemoglobinopathy scheme and MHRA compliance for transfusion should be available. | Y | | Y | |
| HN-401 | **Facilities and Equipment**<br><br>Facilities and equipment should be appropriate for the usual number and case mix of patients. Equipment should be appropriately maintained. | Y | | Y | |
| HN-501 | **Transition Guidelines**<br><br>Guidelines on transition to adult care should be in use covering at least:<br>a. Age guidelines for timing of the transfer<br>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<br>c. Allocation of a named coordinator for the transfer of care<br>d. A preparation period and education programme relating to transfer to adult care<br>e. Communication of clinical information from paediatric to adult services<br>f. Arrangements for monitoring during the time immediately after transfer to adult care<br>g. Arrangements for communication between the Specialist Haemoglobinopathy Centres and Local Haemoglobinopathy Teams | N | Guidelines covering transition were in the process of being agreed.<br>A transition programme was in the process of being implemented using 'Ready Steady Go', and joint clinics were in place with the paediatric service. | N | Guidelines covering transition were in the process of being agreed.<br>A transition programme was in the process of being implemented using 'Ready Steady Go', and joint clinics were in place with the adult service. |

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| HN-502 | **Monitoring Protocols**<br><br>Protocols should be in use covering the monitoring expected at:<br>a. First out-patient appointment (SHC only)<br>b. Routine monitoring<br>c. Annual review (SHC & any LHTs to which annual reviews are delegated)<br>Protocols should cover both clinical practice and information for patients and families. | N | From the evidence seen the annual reviews were incomplete and did not follow the NHR requirements. See also the Further Consideration section of the main report. | N | There was no evidence from the notes seen at the time of the visit that annual reviews took place. |
| HN-503 | **Clinical Guidelines: LHT Management and Referral**<br><br>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 | | N/A | |
| HN-504 | **Transfusion Guidelines**<br><br>Transfusion guidelines should be in use covering:<br>a. Indications for:<br>i. emergency and regular transfusion<br>ii. use of simple or exchange transfusion<br>iii. offering access to automated exchange transfusion to patients on long-term transfusions<br>b. Protocol for carrying out a manual and automated exchange transfusion<br>c. Investigations and vaccinations prior to first transfusion<br>d. Recommended number of cannulation attempts | Y | However, the number of cannulation attempts that should be made by one individual was not explicit in the guidance seen. | N | Reviewers considered that the guidance would benefit from review to be more explicit for the following reasons:<br>c) clarifying the investigations and vaccinations prior to first transfusion; and<br>d) making clear the number of cannulation attempts that should be made by one individual. |

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| HN-505 | **Chelation Therapy**<br>Guidelines on chelation therapy should be in use covering:<br>a. Indications for chelation therapy<br>b. Choice of chelation drug/s, dosage and dosage adjustment<br>c. Monitoring of haemoglobin levels prior to transfusion<br>d. Management and monitoring of iron overload, including management of chelator side effects<br>e. Use of non-invasive estimation of organ-specific iron overloading heart and liver by T2*/R2<br>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.<br>g. Self-administration of medications and infusions and encouraging patient and family involvement in monitoring wherever possible. | Y | | N | Guidelines covering chelation therapy for children and young people were not yet in place. |

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| HN-506 | **Clinical Guidelines: Acute Complications**<br><br>Guidelines on the management of acute complications should be in use covering at least:<br>**For patients with sickle cell disease:**<br>a. Acute pain<br>b. Fever, infection and overwhelming sepsis<br>c. Acute chest syndrome<br>d. Abdominal pain and jaundice<br>e. Acute anaemia<br>f. Stroke and other acute neurological events<br>g. Priapism<br>h. Acute renal failure<br>i. Haematuria<br>j. Acute changes in vision<br>k. Acute splenic sequestration (children only)<br>**For patients with thalassaemia:**<br>l. Fever, infection and overwhelming sepsis<br>m. Cardiac, hepatic or endocrine decompensation | Y | | N | The guidelines did not cover acute complications for patients with thalassaemia. Guidelines were not available covering 'b', 'd', 'h', 'j', 'l' or 'm'. Reviewers considered that the guidance in place for priapism and acute splenic sequestration did not include sufficient detail. |
| HN-507 | **Specialist Management Guidelines**<br><br>Guidelines should be in use covering the care of patients with sickle cell disease and thalassaemia:<br>a. During anaesthesia and surgery<br>b. Who are pregnant<br>c. Receiving hydroxycarbamide therapy | Y | | N | The guidelines seen were out of date and did not cover patients with thalassaemia or the care of children receiving hydroxycarbamide therapy. |

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| HN-508 | **Clinical Guidelines: Chronic complications**<br><br>Guidelines on the management of chronic complications should be in use covering at least:<br>a. Renal disease<br>b. Orthopaedic problems<br>c. Retinopathy<br>d. Cardiological complications / pulmonary hypertension<br>e. Chronic respiratory disease<br>f. Endocrinopathies<br>g. Neurological complications<br>h. Chronic pain<br>i. Liver disease<br>j. Growth delay / delayed puberty (children only)<br>k. Enuresis (children only) | N | Guidelines on the management of chronic respiratory disease, chronic pain and liver disease were not yet in place.<br>Guidance was available for all other aspects of the QS. | N | Guidelines on the management of chronic complications were not yet in place |
| HN-509 | **Referral for Consideration of Bone Marrow Transplantation (Children's Services Only)**<br><br>Guidelines for referral for consideration of bone marrow transplantation should be in use. | N/A | | Y | |
| HN-510 | **Non-Transfusion Dependent Thalassaemia (nTDT)**<br><br>Network-agreed clinical guidelines for the management of Non-Transfusion Dependent Thalassaemia should be in use, covering:<br>a. Indications for transfusion<br>b. Monitoring iron loading<br>c. Indications for splenectomy | Y | | N | Guidelines were not in place covering Non-Transfusion Dependent Thalassaemia. |
| HN-599 | **Clinical Guideline Availability**<br><br>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 | | N | The guidance available on the intranet was either incomplete or out of date. |

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| HN-601 | **Service Organisation**<br><br>A service organisation policy should be in use covering arrangements for:<br>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)<br>b. Ensuring all patients are reviewed by a senior haematology decision-maker within 14 hours of acute admission<br>c. Patient discussion at multi-disciplinary team meetings (QS HN-602)<br>d. Arrangements for liaison with community paediatricians and with schools (children's services only)<br>e. 'Fail-safe' arrangements for ensuring all children and young people have Trans-Cranial Doppler ultrasound when indicated (children's services only)<br>f. Follow up of patients who do not attend<br>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.<br>h. Accessing specialist advice (QS HN-304)<br>i. Two-way communication of patient information between SHC and LHTs<br>j. If applicable, arrangements for coordination of care across hospital sites where key specialties are not located together | N | There was no service organisation policy covering the arrangements defined in the QS. | N | There was no service organisation policy covering the arrangements defined in the QS. |

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| HN-602 | **Multi-Disciplinary Meetings**<br><br>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). | N | Only inpatients were discussed at the general haematology MDT meetings.<br>Arrangements for discussion of other patients were not yet in place. | N | No evidence was available to demonstrate that regular MDT meetings were in place. |
| HN-603 | **Delegation of Annual Reviews**<br><br>If annual reviews are undertaken by LHT/s on behalf of the SHC, a written agreement should be in place covering:<br>a. Monitoring protocols (QS HN-502)<br>b. LHT management and referral guidelines (QS HN-503)<br>c. National Haemoglobinopathy Registry data collection (QS HN-701) | N/A | | N/A | |
| HN-604 | **Out of Hours Elective Care**<br><br>Arrangements should be in place for out of hour's transfusion, phlebotomy and out-patient clinics appropriate to the needs of the local population. | N/A | | N | At the time of the visit the service did not have the capacity to provide out-of-hours elective care. |
| HN-605 | **Service Level Agreement with Community Services**<br><br>A service level agreement for support from community services should be in place covering, at least:<br>a. Role of community service in the care of patients with haemoglobin disorders<br>b. Two-way exchange of information between hospital and community services. | N | The CNS covered the acute and community service. | N | No community service for children and young people with haemoglobin disorders was in place. |

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| HN-606 | **Trans-Cranial Doppler Ultrasound Standard Operating Procedure (Children's Services Only)**<br><br>A Standard Operating Procedure for Trans-Cranial Doppler ultrasound should be in use covering at least:<br>a. Trans-Cranial Doppler modality used<br>b. Identification of ultrasound equipment and maintenance arrangements<br>c. Identification of staff performing Trans-Cranial Doppler ultrasound (QS HN-207)<br>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<br>e. Arrangements for recording and storing images and ensuring availability of images for subsequent review<br>f. Reporting format<br>g. Arrangements for documentation and communication of results<br>h. Internal systems to assure quality, accuracy and verification of results | N/A | | N/A | However, it was not clear to reviewers that a standard operational procedure was in place to cover the visiting service. |
| HN-607 | **Network Review and Learning Meetings**<br><br>At least one representative of the team should attend each Network Review and Learning Meeting (QS HY-798). | N/A | | N/A | |
| HN-608 | **Neonatal Screening Programme Review Meetings**<br><br>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/A | | Y | |

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| HN-701 | **National Haemoglobinopathy Registry**<br><br>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 | Arrangements had been agreed for the SHT to submit data to the NHR in due course. | Y | |
| HN-702 | **Activity Data**<br><br>The service should monitor on an annual basis:<br>a. Number of acute admissions, day unit admissions, Emergency Department attendances and out-patient attendances<br>b. Length of in-patient stays<br>c. Re-admission rate<br>d. 'Did not attend' rate for out-patient appointments | N | Systems to enable access and monitoring of activity data as required by the QS were not yet in place. | Y | |

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| HN-703 | **Quality Dashboard**<br><br>The service should monitor Quality Dashboard data including the number and percentage of:<br>a. Adverse events reported on the NHR for which a mortality or serious case review has taken place<br>b. Children who have had Trans-Cranial Doppler screening undertaken within national guidelines<br>c. Patients given pain relief within half an hour of presentation with sickle crisis<br>d. Patients with possible sickle disorders identified by neonatal screening who have been entered on a care pathway<br>e. Eligible children beginning penicillin at or before three months of age<br>f. Patients registered on the National Haemoglobinopathy Registry who had an annual review undertaken within the last year<br>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<br>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/A | The QS was only applicable for services commissioned by NHS England. | N/A | The QS was only applicable for services commissioned by NHS England. |
| HN-704 | **Other Quality Data**<br><br>The service should monitor on an annual basis:<br>a. Proportion of children who have been offered treatment following Trans-Cranial Doppler Screening | N/A | | Y | |

| Ref | Standard | Adults | | Paediatrics | |
|---|---|---|---|---|---|
| | | Met? | | Met? | Comments |
| HN-705 | **Other Audits**<br><br>Clinical audits covering the following areas should have been undertaken within the last two years:<br>a. Availability of extended red cell phenotype in all patients and the proportion of patients who have developed antibodies<br>b. Whether all eligible patients on long term transfusion have been offered automated exchange transfusion<br>c. Waiting times for elective:<br>i. Phlebotomy<br>ii. Cannulation<br>iii. Setting up of the blood transfusion (for pre-ordered blood) | N | Clinical audits as defined in the QS had not been undertaken in the last two years. | N | Clinical audits as defined in the QS had not been undertaken in the last two years. Audits covering 'time to analgesia' and the numbers of admissions for the sickle cell screening programme had been completed. |
| HN-706 | **Network Audits**<br><br>The service should participate in agreed network-wide audits. | N/A | | N/A | |
| HN-707 | **Research**<br><br>The SHC should actively participate in research relating to the care of patients with haemoglobin disorders. | N | The service was not yet involved in, and did not yet refer patients for inclusion in, research taking place elsewhere relating to the care of patients with haemoglobin disorders. | Y | The service was involved in the HESTIA 3 & 4 research trials. |
| HN-708 | **Trans-Cranial Doppler Quality Assurance (Children's Services Only)**<br><br>The service should monitor and review at least annually:<br>a. Whether all staff performing Trans-Cranial Doppler ultrasound have undertaken 40 procedures in the last year (QS HN-207)<br>b. Results of internal quality assurance systems (QS HN-606)<br>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 | | N/A | Provided by an external provider. |

| Ref | Standard | Adults | | Paediatrics | |
|---|---|---|---|---|---|
| | | Met? | | Met? | Comments |
| HN-798 | **Review and Learning**<br><br>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:<br>a. Review of any patient with a serious adverse event or who died<br>b. Review of any patients requiring admission to a critical care facility<br>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) | N | Mortality and morbidity reviews were in place but not other meetings to discuss the other areas defined in the QS. | N | Mortality and morbidity reviews were in place but not other meetings to discuss the other areas defined in the QS. |
| HN-799 | **Document Control**<br><br>All policies, procedures and guidelines should comply with Trust (or equivalent) document control procedures. | Y | | N | The guidelines in circulation were in draft form and were incomplete. |

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