Health services for people with haemoglobin disorders

University Hospital Southampton NHS Foundation Trust

Visit Date: 28th June 2019

Report Date: October 2019

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Contents

Introduction

This report presents the findings of the peer review of health services for people with haemoglobin disorders provided by the University Hospital Southampton NHS Foundation Trust that took place on 28th June 2019. The purpose of the visit was to review compliance with the Quality Standards for Health Services for People with Haemoglobin Disorders V4, 2018 which were developed by the UK Forum on Haemoglobin Disorders working with the Quality Review Service (QRS). The peer review visit was organised by QRS on behalf of the UK Forum on Haemoglobin Disorders. The Quality Standards refer to the following types of specialised service for people with haemoglobin disorders:

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

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 these proposals. Appendix 1 lists the visiting team that reviewed the services provided by the University Hospital Southampton. Appendix 2 contains the details of compliance with each of the standards and the percentage of standards met.

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

- University Hospital Southampton NHS Foundation Trust
- NHS England Specialised Commissioning – Haemoglobinopathies

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.

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

About the Quality Review Service

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

Expected outcomes are better quality, safety and clinical outcomes, better patient and carer experience, organisations with better information about the quality of clinical services, and organisations with more

confidence and competence in reviewing the quality of clinical services. More detail about the work of QRS is available at www.qualityreviewservicewm.nhs.uk

Acknowledgments

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

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Review Visit Findings

Trust-wide: University Hospital Southampton NHS Foundation Trust

General Comments

This review looked at the care of health services for children, young people and adults with haemoglobin disorders. During the course of the visit, reviewers visited the emergency department (ED), assessment units and wards, and met with patients and carers and with staff providing the haemoglobinopathy service.

University Hospital Southampton served the population in and around Southampton, which was estimated to be 500,000 at around the time of the visit. Patients with complex haemoglobinopathy problems were managed with the support of the adult and paediatric specialist haemoglobinopathy teams.

The prevalence of sickle cell and thalassaemia was relatively low, with approximately 40-50 patients registered with the haemoglobinopathy adult service and 47 patients registered with the haemoglobinopathy paediatric service. Patient numbers fluctuated slightly depending on the number of students with haemoglobin disorders studying at the local university.

The national programme for the reconfiguration of haemoglobinopathy services was ongoing, and the specialist team were not yet clear whether they would be part of the Oxford Haemoglobinopathy Coordinating Centre (HCC) or one of the London HCCs for the care of adults and children with sickle cell disease.

Some comments in the Trust-wide section of this report apply to both the adult or paediatric services and so are not duplicated in other areas of the report.

ADULTS

Trust	Reviewed as:	No. of adults with sickle cell disease	No. of adults with thalassaemia	Usually seen by the linked hospitals/local teams (except for annual reviews)	No. of adults on long-term red cell transfusions
University Hospital Southampton NHS Foundation Trust	SHT	40 - 50	5	36 - 46	8, with fewer than 5 attending Oxford Exchange Transfusion Programme

CHILDREN AND YOUNG PEOPLE

Trust	Reviewed as:	No. of children with sickle cell disease	No. of children with thalassaemia	Usually seen by the linked hospitals/local teams (except for annual reviews)	No. of children on long term red cell transfusions
University Hospital Southampton NHS Foundation Trust	SHT	40	7	30	9

Support groups available for patients and carers	Y/N
Sickle Cell Disease – Adults	N
Thalassemia – Adults	N
Sickle Cell Disease – Children and Young People	N
Thalassemia – Children and Young People	N

Progress since last visit

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

- There had been an increase (from four to eight) in the number of children and young people starting on hydroxycarbamide.
- Four children and young people had been referred for bone marrow transplantation.
- Paediatric guidelines had been agreed with the Southampton and Oxford Retrieval Team (SORT) for children requiring transfer for intensive care support.
- A clinical nurse specialist (CNS) for haemoglobinopathies had commenced in post and was providing acute hospital and community support to patients and their families during normal working hours.
- Patients were being registered onto the National Haemoglobinopathy Register (NHR). Annual reviews for paediatric patients were being submitted to the NHR, and the adult service had plans to enter annual review data for adults.
- Training for ward staff had commenced on the use of patient controlled analgesia (PCA).
- There was an agreement with the adult service based at Guy's and St Thomas' NHS Foundation Trust (GSTT) to provide joint clinics.
- The adult service had received confirmation of Specialist Haemoglobinopathy Team (SHT) status as part of the national reorganisation of haemoglobinopathy services.

Immediate Risk:

1 **Guidelines**

The guidelines covering most aspects of acute care of people with haemoglobin disorders (adults and children) either were not yet agreed or did not include the latest national best practice guidance.

The lack of agreed and robust clinical guidelines had been identified as an immediate risk following the last peer review visit in 2015. The UK Forum on Haemoglobin Disorders validation panel had also given feedback to the team in March 2019 following the validation of the adult services self-assessment. This feedback suggested that the team should ensure that the draft Trust guidelines in use (which were adapted from the GSTT clinical guidelines for sickle cell disease of 2011) reflected the latest guidance available.

In the view of the reviewers, the lack of agreed and robust clinical guidelines at the time of the visit was an immediate risk to patient safety and clinical outcomes because:

a. The draft guidelines covering the care of people with haemoglobin disorders (adults), which were in the process of being agreed, were not up to date; for example:

   i. Indications for drug therapy for patients on hydroxycarbamide were not consistent with the British Society for Haematology Guidance (2018).

   ii. The guidance for use of deferasirox included the use of soluble tablets, which are no longer available, and therefore the dosages documented did not correlate with the doses of enteric coated preparations that are now widely used.

b. The guidelines covering the care of people with haemoglobin disorders (children), which were accessible on the Trust intranet, were out of date; for example:

   i. Prescribing of hyperhydration for patients with sickle cell disease, and the use of diuretics in children with acute chest syndrome, is no longer recommended.

   ii. Taking arterial blood gas as part of the monitoring of children who are acutely unwell is not deemed best practice.

   iii. There were discrepancies in the exchange transfusion target, with the target being stated as both ‘less than 20%’ and ‘less than 25-35%’.

   iv. Haemoglobin concentration was expressed as gram/decilitre rather than the standard measurement of gram/litre, which has the potential for confusion.

c. The Trust Adult Blood Transfusion Policy (2018) did not include any specific guidance on transfusing patients with sickle cell disease and thalassaemia. In particular, there was no guidance on manual and automated exchange transfusions, and the need for extended phenotype matching prior to transfusion was not mentioned.

Reviewers were also extremely concerned that a robust governance process was not in place for the development and agreement of clinical guidelines within the Trust. Staff who met with the reviewers commented that the Trust governance process was lengthy, and that the clinical guidelines for adults had been in the process of ratification for 18 months.

Reviewers considered that it was important that all staff were able to access comprehensive and evidence-based guidance at all times. Because of the low numbers of patients with haemoglobin disorders cared for by the Trust, some staff may be less familiar with patients with haemoglobin disorders, which further increased the risk arising from the lack of availability of robust clinical guidelines. As a matter of urgency, whilst a comprehensive suite of guidance is agreed, the Trust should ensure that guidance covering at least acute chest syndrome, manual and automated exchange transfusions and acute pain

management for patients with haemoglobin disorders is available to staff. Liaising with, and adopting guidelines from, another specialist centre should be considered in the interim, as this would ensure that appropriate guidance was available quickly. A clear Trust policy that links to the relevant external guidelines was highly recommended by reviewers[1].

Good Practice

1. Reviewers were impressed with the work that the CNS had achieved since commencing in post after the last visit. Liaison with the community children's service had improved, enabling patients to have hydroxycarbamide blood monitoring in the community, and the CNS was working with schools to ensure education and care plans were implemented, supporting young people to transition from the paediatric to adult service and liaising with local referring teams. The CNS had also undertaken training in delivering apheresis and non-medical blood prescribing. The CNS had implemented education sessions for ward staff covering haemoglobin disorders and patient controlled analgesia (PCA).
2. The reviewers acknowledged the work undertaken by the University Hospital Southampton NHS Foundation Trust in developing the 'Ready Steady Go' programme for young people to transition to adult services which had been adopted for use nationally.

[1] **Trust response to Immediate Risk:** We have an action plan in place to make available the full guidelines following expedited review of the adult sickle cell guidelines. We have been in contact with the lead commissioner and are looking to secure a date for a meeting in the week commencing 22 July 19 to discuss the QRS Review of Health Services for People with Haemoglobin Disorders Immediate Risk in more detail. In the interim the following steps have been taken:
The Clinical Pathways Review Group (CPRG) met on 9th July 2019 and reviewed the existing version of the guidelines that have been in the review process for some time. Specific requests regarding use of abbreviations and consistency within the guidelines have been made as well requesting further medicines committee and radiology review. The guidelines will be reviewed at the medicines committee on 17th July 2019 and we will incorporate their changes in to the current version and I have sought agreement from the chair of the CPRG that he will review the amended guidelines by 30th July 2019. In relation to the specific points raised in your letter, we propose the following actions:
1a) Adult guidelines: (i) Indications for hydroxycarbamide will be updated in line with BSH (2018) guidance. (ii) Guidance for deferasirox will be amended to reflect the current formulation. 1b) Paediatric guidelines: (i) Reference to hyperhydration and diuretics will be removed. (ii) Use of arterial blood gases for monitoring unwell children will be removed. (iii) Guidance for target values for exchange transfusion will be unified between sections. (iv) Haemoglobin units will be changed to g/L .
1c) Blood transfusion policy:
This will be updated to include specific reference to transfusing people with sickle cell anaemia and thalassaemia including the use of manual and automated exchange transfusions and the requirement for extended phenotyping. We fully recognise that the lengthy review process for agreement of clinical guidelines within the Trust has resulted in sections of the guidelines becoming outdated. Once the current guidelines are approved and made available, we propose to separate the two sets of adult and paediatric guidelines in to a series of individual guidelines. This will enable updates and amendments to take place to individual sections in a more timely manner. We have agreed with NHS England that additional resource will be made available to UHS to fulfil our responsibilities as a Specialist Haemoglobinopathy Centre and this investment is based on the understanding that the £107,500 funding for being recognised as a Specialist Haemoglobinopathy Team (SHT) is an annual payment for the minimum of the next 3 years. This will require significant job plan and staffing reorganisation and potentially appointments therefore I propose that this is undertaken and completed by 30th September 2020. Some sections will be dependent on designation of our Regional Haemoglobinopathy Co-ordinating Centre as we will need to align our guidelines with that centre. We propose that the guidelines will be updated on an annual basis and as needed based on new evidence in association with the RCC via a process agreed with that centre. In order to mitigate against significant potential clinical risks, the following adult guidelines have been made available on the Hospital 'staffnet' (intranet) on the Clinical Haematology pages on 5th July 2019. These are accessible by all staff in the Trust: Acute chest syndrome Red cell exchange transfusion and Acute pain management for patients with haemoglobin disorders
WMQRS Response to Trust actions: We have now considered your response and can confirm that the actions as described will address the immediate risk identified during the visit, once fully implemented.

Concerns

1. **Emergency Department Training**

   A programme of training for ED staff covering haemoglobin disorders was not yet in place to ensure that staff were aware of the needs of patients with haemoglobin disorders attending the department in an emergency. Patients who met with the visiting team also considered that, when they attended the ED, staff in the department did not have sufficient understanding of haemoglobin disorders and the need to give appropriate and timely analgesia.

2. **Access to Analgesia**

   Patients who met with the visiting team commented that they did not always receive timely access to appropriate analgesia if they were admitted to other wards. As a number of staff on Ward D2 who had completed the training on PCA were no longer in post, access to PCA for patients admitted to Ward D2 was not always possible. Patients who were admitted to non-haematology outlying wards were not able to use this method of analgesia.

3. **Access to Psychology**

   Access to psychology was limited, and none of the patients who met with the visiting team were aware of the support available. In practice, referrals could be made to the general psychology service, and the ward staff who met with the visiting team would refer patients to the palliative care team for support. However, neither of these services would have the relevant experience in caring for patients and families with haemoglobin disorders. Reviewers were told that psychological support was available from the visiting team from GSTT when they attended the joint clinic, but a joint clinic had not taken place for over 12 months.

4. **Clinical Nurse Specialist**

   At the time of the visit there was no cover available for absences of the CNS, who was providing acute hospital and community support to children, young people and adults.

Further Consideration

1. The transition process was not yet fully integrated, and there was no clear process for transitioning into adult care. Reviewers met with a number of parents and young adults who had not had the option of completing a formal transition process prior to being seen in the adult service. Reviewers were told that the lead nurse was now leading on the coordination of the transition pathway across both the adult and the paediatric services, and that the process being implemented would ensure that all young people would be given the appropriate information and support to transition to adult care.
2. Access to neuro-psychology testing was not yet in place, particularly for children, to assess any early signs of cognitive impairment and those who were at high risk of a stroke. Reviewers considered that there may be opportunities to work with a clinical psychologist with specific training in this area, such as the Trust stroke service or local education providers.
3. As part of the work to develop the service, identifying 'link' nurses in both the acute and community settings may help with building a wider network of expertise within the Trust and externally, especially with local referring teams.
4. Reviewers expressed concern at the over-reliance on the CNS to provide a wide range of services. The CNS covered the acute hospital, led on transition and education for staff and provided support in the community, as well as being the lead in many service developments such as guideline development. The CNS was also spending time on administration and data collection, which reviewers considered was not

an appropriate use of clinical time. Reviewers considered that the role would not be sustainable in the future without additional clinical leadership and support.

5. Patients were registered on National Haemoglobinopathy Register, but only a limited amount of other data were entered, because of the lack of data management support.

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Specialist Haemoglobinopathy Team (Adult Services): University Southampton Hospital NHS Foundation Trust

General Comments and Achievements

The service was led by a consultant haematologist and CNS, and patients and carers who met with the visiting team were appreciative of the care they received from the lead team. There was evidence of collaborative working and good relationships with both the managerial team and the paediatric service. Good support was available from the Trust acute pain team for inpatients. The facilities available within the haematology day unit and ward were welcoming and of a high quality.

The level of training for Ward D2 staff had improved since the last visit. In addition to training for staff on the use of PCA, four of the ward staff had attended training on the care of patients with sickle cell disease, and another session was due to be run in September for a further five ward staff.

Out of the forty patients cared for by the team, twelve patients were taking hydroxycarbamide regularly and five were taking regular iron chelation, with one of these being on dual therapy. Three thalassaemia patients were under the care of the Whittington Health NHS Trust for follow up and annual review.

Inpatient admissions were low, with approximately one to two inpatient admissions to the hospital (Ward D2) each month.

The vision to develop the apheresis service to offer automated exchange transfusions, which was noted at the last visit in 2015, had not materialised, but the team were hopeful that this service could be implemented in the near future.

The report contains a number of issues that would need to be addressed in order for the team to function as a comprehensive Specialist Haemoglobinopathy Team (SHT).

<table>
<tr><th>Specialist Haemoglobinopathy Centre</th><th>Links to Local Haemoglobinopathy Teams</th></tr>
<tr><td rowspan="3">University Hospital Southampton NHS Foundation Trust</td><td>• Dorset County Hospital NHS Foundation Trust
• Hampshire Hospitals NHS Foundation Trust – Basingstoke and North Hampshire Hospital
• Portsmouth Hospitals NHS Trust
• Isle of Wight NHS Trust – St Mary’s Hospital
• Poole Hospital NHS Foundation Trust
• Royal Hampshire County Hospital
• The Royal Bournemouth and Christchurch Hospitals NHS Foundation Trust
• Salisbury NHS Foundation Trust
• Western Sussex Hospitals NHS Foundation Trust – St Richard’s Hospital</td></tr>
<tr><td>Links to Specialist Haemoglobinopathy Teams</td></tr>
<tr><td>• Oxford University Hospitals NHS Foundation Trust (exchange transfusion programmes)
• The Whittington Health NHS Trust (thalassaemia patients annual review)
• Guy’s and St Thomas’ NHS Foundation Trust</td></tr>
</table>

Staffing

Staffing for the SHT Adult Haemoglobinopathy Service [2]	Number of patients	Actual WTE (at time of the visit)
Consultant haematologist - >0.6 WTE per 150 patients dedicated to work with patients with HBO	45-55	0.1
At least 0.25 WTE allocated to haemoglobinopathies CPD in the Adult Consultant Job plan	45-55	0
Trust employ a clinical psychologist for adult patients who has >0.5 WTE per 200 patients dedicated to work with patients with HBO?	45-55	0

Emergency Care:

Patients attending the ED were triaged and during normal working hours the lead consultant and nurse were generally notified. Care plans were accessible via the electronic patient records system.

Inpatient Care:

The inpatient ward (D2) was a 20-bedded dedicated haematology/oncology unit. It had six rooms with en-suite facilities and two bays of five beds, one bay with four beds, with an en-suite in each bay. Occasionally patients were admitted to other wards within the hospital.

Day unit Care:

Patients who required maintenance blood transfusions and manual exchange transfusions would attend the haematology day unit (C7); this had extended opening hours of 8am to 8pm on weekdays and 8am to 4pm on Saturdays. The day unit was run by trained nursing staff, with support from junior and middle grade medical staff and the haematology consultants.

Outpatient Care:

A specialist haemoglobinopathy outpatient clinic was held on the first Thursday afternoon of each month and was led by the lead clinician and CNS.

Community-based Care:

The haemoglobinopathy community nurse specialist provided some community care for adult patients, and the CNS had good links with other community services.

Views of Service Users and Carers

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

- Overall, the users and carers were highly complementary about the care and support available from the lead clinician and CNS.
- The users and carers who met with the visiting team were enthusiastic about having the opportunity to meet other patients; in particular, the younger adults were interested in meeting those older than themselves. Their enthusiasm was such that it led to the development of a social media group by the end of the meeting, and there were plans to widen the invitation to other patients in order to formalise a support group for all patients with haemoglobin disorders.

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

- The CNS would email a patient's care plan to them, which the patients found useful, especially as they could share it with other health professionals involved in their care and in emergency situations. The patients were not aware of the use of 'emergency cards', which they thought would be useful.
- In an emergency, the users and carers were not confident that staff in the emergency department had sufficient understanding of haemoglobin disorders, commenting that access to analgesia was often difficult and that there was an initial reluctance by staff to check their emergency care plan on the electronic reports system. Similar concerns were expressed about admission to wards, other than Ward D2, across the Trust.
- The young people who met with the visiting team had not had information or formal preparation prior to their transition to the adult service, although one user was aware of the 'Ready Steady Go' programme that had been devised by the Trust.

Good Practice

1. In 2018 one consultant had been recognised by the Sickle Cell Society for their work with patients with sickle cell disease See also trust wide section of the report

Immediate Risks: See the Trust wide section of the report

Serious Concern

1. **Ward nurse competences**

   From discussion with staff, reviewers were not assured that staff would recognise acute complications for patients with haemoglobin disorders such as acute chest syndrome. Staff on Ward D2 had completed some training on haemoglobin disorders, but this training was not based on a competence framework.

   Staff commented that the number of patients admitted to the ward was low, and that maintaining competences was difficult. Reviewers considered that developing a template nursing care plan with prompts to help recognise the acutely unwell patient with haemoglobin disorders may be helpful.

Concern

1. **Consultant staffing**

   Reviewers were concerned about whether the service had sufficient consultant medical staff with appropriate competences in the care of people with haemoglobin disorders to provide staffing for regular reviews, emergency care and clinics. At the time of the visit, the consultant had only one programmed activity session (PA) for work with the service, and was also advising the antenatal screening service. Cover for the lead consultant was from other consultant haematologists who might not have competences in haemoglobin disorders, and therefore it was not always possible to ensure that all patients were reviewed by a senior haematology decision-maker (with appropriate competences) within 14 hours of acute admission.

   Reviewers commented that it may not be easy to increase specialist consultant support because of the low numbers of patients, and therefore that joint support with other centres may be an option.

2. **Multidisciplinary discussion**

   Multidisciplinary team (MDT) meetings to discuss and review patient care were not yet in place. Inpatients were discussed as part of the wider weekly haematology MDT, but there was no formal mechanism for multidisciplinary discussion of complex and community-based patients or with other local referring teams.

3. **Service operational policy**

   No service operational policy was in place covering the clinical and managerial aspects of the service. This issue had also been identified during the previous review.

4. **Patient information**

   The iron chelation information was out of date, and included information for patients about having a liver biopsy rather than an MRI scan. The written information about the long-term use of hydroxycarbamide would benefit from review to ensure that sexual health, the possible side effects of long-term therapy, and fertility, including sperm banking, are clearly explained. Reviewers considered that this would be particularly important for young people who are transitioning to adult care, to ensure that they have sufficient knowledge to give informed consent.

5. See also Trust-wide concerns section of the report.

Further Consideration

1. The clinic letters seen by the reviewers were not clear about monitoring of patients. The letters were clear about the actions taken at each review, and in practice monitoring was appropriate, but the documentation was not specific about future monitoring requirements, which would make it difficult if other staff were accessing the patients' notes to see what monitoring was required.
2. Little patient information was available for patients with thalassemia. Staff who met with the visiting team commented that this was because Southampton was a low prevalence area and patients were seen individually. Reviewers considered that, regardless of the number of patients being cared for by the team, localised information should be available.
3. The care plans that were held electronically were not 'tagged', making it more difficult to navigate on the patient record system.
4. See also Trust-wide further consideration section of the report.

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Specialist Haemoglobinopathy Team (Children and Young People Services): University Hospital Southampton NHS Foundation Trust

General comments

The service was led by a consultant paediatric haematologist and CNS and parents and carers who met with the visiting team were appreciative of the care they received from the lead team. There was evidence of collaborative working and good relationships with both the managerial team and the adult service. As a Specialist Haemoglobinopathy Team (SHT), the paediatric team provided care to local patients and delivered annual reviews for approximately 30 children and young people living in the Wessex area, and it also provided cover for the Isle of Wight and the Channel Islands.

Specialist Haemoglobinopathy Centre	Local Haemoglobinopathy Teams
University Hospital Southampton NHS Foundation Trust	• Portsmouth Hospitals NHS Trust • Poole Hospital NHS Foundation Trust • Salisbury NHS Foundation Trust • Western Sussex Hospitals NHS Foundation Trust – St Richards Hospital Chichester • Royal Hampshire County Hospital • Hampshire Hospitals NHS Foundation Trust – Basingstoke and North Hampshire Hospital

Staffing

Staffing for the SHT Paediatric Haemoglobinopathy Service [3]	Number of patients	Actual WTE (at time of the visit)
Consultant haematologist/paediatrician - >0.6 WTE per 150 patients dedicated to work with patients with HBO	47	0.1
Is at least 0.25 WTE allocated to haemoglobinopathies CPD in the Paediatric Consultant Job	47	0
Trust employ a clinical psychologist for paediatric patients who has >0.5 WTE per 150 patients dedicated to work with patients with HBO ?	47	0

Emergency Care:

Very few children and young people attended the paediatric ED, because they had open access to the paediatric unit. Parents who met with the visiting team were all aware of the process for contacting the unit when required.

Ward Care:

Patients requiring acute or elective admission were admitted to the paediatric medical unit.

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

Day unit Care:

Day care was provided on the John Atwell Day Unit, which had extended opening hours from 7am to 8pm on weekdays. Children and young people were admitted for day case management and for top-up transfusions. Children and young people who required automated exchange transfusions had to attend the haemodialysis unit (G4). All nurses had competences in phlebotomy and cannulation.

Outpatient Care:

Paediatric specialist haemoglobinopathy clinics were held on the fourth Tuesday of each month and were led by the consultant haematology paediatrician and the CNS. A radiographer trained to perform transcranial Doppler ultrasound was also available to attend each clinic. Children who were diagnosed with sickle cell disease from the neonatal screening programme were referred directly to the lead consultant and seen by the team in the monthly clinics.

Community-based Care:

The haemoglobinopathy community nurse specialist provided some community care for paediatric patients, undertook visits to schools and was involved in the development of health and education care plans for children with haemoglobin disorders. The CNS had good links with the community children's nurses who would take pre-transfusion bloods for patients, and bloods for patients on hydroxycarbamide.

Views of Service Users and Carers

The visiting team met with four parents during the course of the visit. Overall, the parents were highly complimentary about the care their children received. In addition:

- Parents spoke consistently about the enormous amount of support and guidance they received from the lead consultant and CNS.
- Parents commented that support was always available from the CNS if they raised issues relating to their child's education or school. The CNS was quick to meet with children, their parents and school staff to ensure that problems were quickly resolved.
- Parents were particularly grateful to the CNS, who they would contact as their first point of call for any issues, as they considered that their GPs were not as knowledgeable as the CNS about sickle cell disease.

Good Practice

1. The environment on the paediatric ward was very welcoming, with a new paediatric ED since the last visit. The reviewers were particularly impressed with the open play area. Feedback from patients and carers was actively sought, and a clear graphical display of patient views was noted in ward areas.
2. Reviewers were impressed with the paediatric clinic letters seen at the visit. The letters were very comprehensive, with clear guidance documented for children, parents and GPs.
3. The paediatric section of the intranet 'Pier' provided a good resource for staff. The website was very clear and intuitive.
4. Good links with the community children's team were in place. The community team provided a paediatric phlebotomy service, which reduced the number of times children and parents needed to attend the hospital for drug therapy monitoring and before a blood transfusion.
5. The local referring team from Portsmouth was very appreciative of the support and advice from the CNS for patients with complex needs.
6. See also Trust-wide good practice section of the report.

Immediate Risks: See the Trust-wide section of the report.

Concern

1. **Paediatric Consultant Staffing**

   Reviewers were concerned about whether the service had sufficient paediatric consultant medical staff with appropriate competences in the care of people with haemoglobin disorders to provide staffing for clinics, regular reviews and emergency care. At the time of the visit, the consultant had only one programmed activity session (PA) for work with the service. Cover for the lead consultant was from the lead adult haemoglobin disorders consultant or from the other adult haematologists, who might not have competences in caring for children and young people with haemoglobin disorders.

2. See also Trust-wide concerns section of the report.

Further Consideration

1. See Trust-wide further consideration section of the report.

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Commissioning

General Comments

There was good specialised commissioning engagement with the Trust's recent and successful application to function as a Specialist Haemoglobinopathy Team. The review team met the regional NHSE commissioner who was keen to work with the Trust in order to ensure that high quality clinical care is provided to patients, including those attending local hospitals within the region.

The Immediate Risk issues in the provider section of this report will also require commissioner monitoring and support to ensure they are addressed:

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APPENDIX 1 Membership of Visiting Team

Visiting Team		
Jacqui Bowyer	Paediatric Nurse Specialist-Community	London North West University Healthcare NHS Trust
Dr Subarna Chakravorty	Consultant Paediatric Haematologist	King's College Hospital NHS Foundation Trust
Dr Arne de Kreuk	Consultant Haematologist	North Middlesex University Hospital NHS Trust
Sandy Hayes	Adult Haemoglobinopathy Senior Specialist Nurse and Thames Valley Network Lead Nurse	Oxford University Hospitals NHS Foundation Trust
Roanna Maharaj	Patient Representative	UK Thalassaemia Society
Rachel McFee	Manager of OSCAR	OSCAR Patients Group, Sandwell
Rita Protopapa	Quality Assurance Programme Manager (Haematology)	St George's University Hospitals NHS Foundation Trust
Dr Tullie Yeghen	Consultant Haematologist	Lewisham and Greenwich NHS 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

Service	Number of Applicable QS	Number of QS Met	% met
Adults- SHT	42	9	21
Children - SHT	50	16	32
Total	**92**	**25**	**27**

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Specialist Services for People with Haemoglobin Disorders

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	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)	N	Generic patient information about the Trust services was available but limited information about the haemoglobinopathy service. Information was not available covering:- 'c', Information about the ward 'e', Explaining about the community service (provided by the Trust). 'g' where to go in an emergency 'h - i-iii'	N	Generic patient information about the Trust services was available but limited information about the haemoglobinopathy service. Information was not available covering:- 'c', Information about the ward 'e', Explaining about the community service (provided by the Trust). 'g' where to go in an emergency 'h - i-iii' . A Sickle Cell Disease handbook for parents was accessible but did not include any local information.

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
HN-102	**Information about Haemoglobin Disorders** Written information should be offered to patients and, where appropriate, their carers covering at least: a. A description of their condition (SC or T), how it might affect them and treatment available b. Problems, symptoms and signs for which emergency advice should be sought c. How to manage pain at home (SC only) d. Splenic palpation and Trans-Cranial Doppler scanning (children only) e. Transfusion and iron chelation f. Possible complications, including priapism and complications during pregnancy g. Health promotion, including: i. Travel advice ii. Vaccination advice h. National Haemoglobinopathy Registry, its purpose and benefits i. Self-administration of medications and infusions	N	Limited information covering the requirements of the QS was seen for patients with Thalassaemia. Information was not available about blood transfusions and the iron chelation information was out of date and did not cover side effects and monitoring. The patient information for hydroxycarbamide did not cover side effects including contraception and sexual health.	N	Limited information covering the requirements of the QS was seen for patients with Thalassaemia. Information was not available about blood transfusions and the iron chelation information was out of date and did not cover side effects and monitoring. The patient information for hydroxycarbamide did not cover side effects including contraception and sexual health.

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	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	Two of the patients who met with the reviewing team commented that the CNS would email their care plan. However, other patients were not clear that they received a copy of their care plan or written summary of their annual review.	Y	
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 for 'c iii' or 'd' was not included in the examples of communication to the Primary Health Care Team seen during the visit.	N	Information for 'c iii' or 'd' was not included in the examples of communication to the Primary Health Care Team seen during the visit.

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
HN-105	**Information about Trans-Cranial Doppler Ultrasound (Children's Services Only)** Written information should be offered to patients and their carers covering: a. Reason for the scan and information about the procedure b. Details of where and when the scan will take place and how to change an appointment c. Any side effects d. Informing staff if the child is unwell or has been unwell in the last week e. How, when and by whom results will be communicated	N/A		Y	
HN-106	**School Care Plan (Children's Services Only)** A School Care Plan should be agreed for each child or young person covering, at least: a. School attended b. Medication, including arrangements for giving / supervising medication by school staff c. What to do in an emergency whilst in school d. Arrangements for liaison with the school e. Specific health or education need (if any)	N/A		Y	The Lead Nurse, if invited, would also attend schools.
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		Y	

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
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	Formal transition arrangements had not yet been fully implemented. Reviewers met with five young people during the course of the visit none of whom had been offered a formal transition process. One patient was aware of the 'Ready, Steady, Go' approach but had not been offered this prior to transfer to the adult service.	N	Formal transition arrangements had not yet been fully implemented. Reviewers met with young people and carers who were aware that they would transition to the adult service, but had not all been offered all the requirements of the QS.
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	N	Mechanisms for involving patients about the organisation of the service was not yet in place. Patient surveys were undertaken but not evidence of changes made as a result of feedback.	N	Mechanisms for involving patients about the organisation of the service was not yet in place. Patient surveys were undertaken but not evidence of changes made as a result of feedback.

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	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 1 PA allocated to the haemoglobinopathy service (clinical, administration and geographical lead).	N	The lead Consultant only had 1 PA allocated to the haemoglobinopathy service (clinical, administration and geographical
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 did not have cover for absences. All other aspects of the QS were met.	N	The lead nurse did not have cover for absences. All other aspects of the QS were met.

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
HN-203	**Medical Staffing and Competences** The service should have sufficient medical staff with appropriate competences in the care of people with haemoglobin disorders, including: **All services:** a. Haematology or paediatric medical staffing for clinics and regular reviews b. 24/7 consultant and junior staffing for emergency care **SHTs only:** c. A consultant specialising in the care of people with haemoglobin disorders on call and available to see patients during normal working hours d. If doctors in training are part of achieving 'a' or 'b' then they should have the opportunity to gain competences in all aspects of the care of people with haemoglobin disorders Staffing levels should be appropriate for the number of patients cared for by the service and its role in the network (SHT/LHT). Staff working with children and young people should have competences in caring for children as well as in haemoglobin disorders. Cover for absences should be available.	N	The lead Consultant only had 1 PA allocated to the haemoglobinopathy service (clinical, administration and geographical lead). 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 .	N	The lead Consultant only had 1 PA allocated to the haemoglobinopathy service (clinical, administration and geographical lead).Haematology cover for the lead clinician was from the adult haematology consultants.

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
HN-204	**Nurse Staffing and Competences** The service should have sufficient nursing staff with appropriate competences in the care of people with haemoglobin disorders, including: a. Clinical nurse specialist/s with responsibility for the acute service b. Clinical nurse specialist/s with responsibility for the community service c. Ward-based nursing staff d. Day unit (or equivalent) nursing staff e. Nurses with competences in cannulation and transfusion available at all times patients attend for transfusion Staffing levels should be appropriate for the number of patients cared for by the service and its role in the network (SHT/LHT). Staff working with children and young people should have competences in caring for children as well as in haemoglobin disorders. Cover for absences should be available.	N	The CNS did not have cover for absences. There was no evidence that ward staff had completed competences in haemoglobin disorders. 'a and b' were met in terms of competences but not the CNS did not have any cover for absences. Reviewers suggested that developing link nurses with an interest in haemoglobin disorders in both the acute and community services may be helpful.	N	The CNS did not have cover for absences. All other aspects of the QS were met.

		Adults- SHT		Children - SHT	
Ref	**Standard**	**Met?**	**Comments**	**Met?**	**Comments**
HN-205	**Psychology Staffing and Competences** The service should have sufficient psychology staff with appropriate competences in the care of people with haemoglobin disorders, including: a. An appropriate number of regular clinical session/s for work with people with haemoglobin disorders and for liaison with other services about their care b. Time for input to the service's multi-disciplinary discussions and governance activities c. Provision of, or arrangements for liaison with and referral to, neuro-psychology Staffing levels should be appropriate for the number of patients cared for by the service and its role in the network (SHT/LHT). Staff working with children and young people should have competences in caring for children as well as in haemoglobin disorders. Cover for absences should be available.	N	No specific psychology support for people with haemoglobin disorders was available and patients were not aware that they could access support from the generic psychology service. Reviewers were told that psychology support was accessible from the 'London team' when providing a visiting clinic, but this clinic had not taken place for over 12 months. *(British Psychology Society Special Interest Group for Psychologists working in Sickle cell and Thalassaemia (2017) suggests 1w.t.e. for 300 patients).*	N	No specific psychology support for people with haemoglobin disorders was available and patients were not aware that they could access support from the generic psychology service. *(British Psychology Society Special Interest Group for Psychologists working in Sickle cell and Thalassaemia (2017) suggests 1w.t.e. for 300 patients).*
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 for all staff was not yet in place. The Lead Nurse did have competences in automated exchange transfusion and blood prescribing.	N	A training plan for all staff was not yet in place. The Lead Nurse did have competences in automated exchange transfusion and blood prescribing.

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	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.	N/A		Y	
HN-299	**Administrative, Clerical and Data Collection Support** Administrative, clerical and data collection support should be appropriate for the number of patients cared for by the service.	N	No admin or clerical support was available to help with data management.	N	No admin or clerical support was available to help with data management.
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	Referrals could be made to all the support services as required by the QS.	Y	Referrals could be made to all the support services as required by the QS .

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
HN-302	**Emergency Department – Staff Competences** Medical and nursing staff working in the Emergency Department/s should have competences in urgent care of people with haemoglobin disorders.	N	A training programme for ED staff was not yet in place and several of the patients who met with the reviewing team commented that staff in the ED did not understand about haemoglobin disorders, especially about the need for giving appropriate analgesia within 30 mins of arrival.	N	A training programme for ED staff was not yet in place and several of the patients who met with the reviewing team commented that staff in the ED did not understand about haemoglobin disorders, especially about the need for giving appropriate analgesia within 30 mins of arrival.
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	N	Patients requiring erythrocytapheresis were referred to Oxford. Manual exchange transfusions were only available if the lead clinician was on site otherwise junior staff would need to contact the lead clinician for advice.	Y	

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

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
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		Y	
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		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	Guidelines were in place but see comments in the main report about feedback from patients.	Y	Guidelines were in place but see comments in the main report about feedback from patients.

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
HN-502	**Monitoring Protocols** Protocols should be in use covering the monitoring expected at: a. First out-patient appointment (SHT only) b. Routine monitoring c. Annual review (SHT & any LHTs to which annual reviews are delegated) Protocols should cover both clinical practice and information for patients and families.	N	Monitoring protocols covering all the requirements of the QS were not yet agreed. However, from the evidence seen, the patient letters did include the relevant information from the first out-patient appointment.	Y	A standard operational procedure was in place and patient letters seen did include the relevant information from the first out-patient appointment
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		N/A	
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	N	The Trust-wide transfusion guidelines did not include specific guidance for transfusing patients with haemoglobin disorders, especially for those who required any exchange transfusion. See the immediate risk section of the report	N	The Trust-wide transfusion guidelines did not include specific guidance for transfusing patients with haemoglobin disorders, especially for those who required any exchange transfusion. See the immediate risk section of the report

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
HN-505	**Chelation Therapy** Guidelines on chelation therapy should be in use covering: a. Indications for chelation therapy b. Choice of chelation drug/s, dosage and dosage adjustment c. Monitoring of haemoglobin levels prior to transfusion d. Management and monitoring of iron overload, including management of chelator side effects e. Use of non-invasive estimation of organ-specific iron overloading heart and liver by T2*/R2 f. Where prescribing is undertaken through shared care arrangements with GPs, guidelines for GPs on prescribing, monitoring and indications for seeking advice from and referral back to the LHT/SHT. g. Self-administration of medications and infusions and encouraging patient and family involvement in monitoring wherever possible.	N	Up to date guidelines were not in use. See immediate risk section of the report	N	Up to date guidelines were not in use. See the immediate risk section of the report

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
HN-506	**Clinical Guidelines: Acute Complications** Guidelines on the management of acute complications should be in use covering at least: **For patients with sickle cell disease:** a. Acute pain b. Fever, infection and overwhelming sepsis c. Acute chest syndrome d. Abdominal pain and jaundice e. Acute anaemia f. Stroke and other acute neurological events g. Priapism h. Acute renal failure i. Haematuria j. Acute changes in vision k. Acute splenic sequestration (children only) **For patients with thalassaemia:** l. Fever, infection and overwhelming sepsis m. Cardiac, hepatic or endocrine decompensation	N	Guidelines had not yet been agreed. The draft guidelines that were awaiting ratification were out of date, and reviewers were told that the process for agreeing the guidelines within the Trust Governance process had been ongoing for the last 18 months. See the Immediate Risk section of the report.	N	The guidelines availability on the Trust intranet were out of date. See the immediate risk section of the report.
HN-507	**Specialist Management Guidelines** Guidelines should be in use covering the care of patients with sickle cell disease and thalassaemia: a. During anaesthesia and surgery b. Who are pregnant c. Receiving hydroxycarbamide therapy	N	As QS -506. Reviewers were concerned that guidance had not yet been agreed as women were being seen in a joint obstetric clinic.	N	As QS -506.

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
HN-508	**Clinical Guidelines: Chronic complications** Guidelines on the management of chronic complications should be in use covering at least: a. Renal disease b. Orthopaedic problems c. Retinopathy d. Cardiological complications / pulmonary hypertension e. Chronic respiratory disease f. Endocrinopathies g. Neurological complications h. Chronic pain i. Liver disease j. Growth delay / delayed puberty (children only) k. Enuresis (children only)	N	As QS -506.	N	As QS -506.
HN-509	**Referral for Consideration of Bone Marrow Transplantation (Children's Services Only)** Guidelines for referral for consideration of bone marrow transplantation should be in use.	N/A		Y	Several children had undergone bone marrow transplantation
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	N	No guidelines had been agreed covering the requirements of the QS	N	No guidelines had been agreed covering the requirements of the QS

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
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.	N	As QS -506	N	The guidelines seen by the reviewer were out of date. See main report.

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
HN-601	**Service Organisation** A service organisation policy should be in use covering arrangements for: a. 'Fail-safe' arrangements for ensuring all children with significant haemoglobinopathy disorders who have been identified through screening programmes are followed up by a SHT (Children's SHT only) b. Ensuring all patients are reviewed by a senior haematology decision-maker within 14 hours of acute admission c. Patient discussion at multi-disciplinary team meetings (QS HN-602) d. Arrangements for liaison with community paediatricians and with schools (children's services only) e. 'Fail-safe' arrangements for ensuring all children and young people have Trans-Cranial Doppler ultrasound when indicated (children's services only) f. Follow up of patients who do not attend g. Transfer of care of patients who move to another area, including communication with all SHT, LHTs and community services involved with their care before the move and communication and transfer of clinical information to the SHT, LHT and community services who will be taking over their care. h. Accessing specialist advice (QS HN-304) i. Two-way communication of patient information between SHT and LHTs j. If applicable, arrangements for coordination of care across hospital sites where key specialties are not located together	N	There was no service organisation policy. For patients who required admission, staff would call the on-call haematologist. At weekends and out of hours there would be a delay in patients being reviewed by a senior haematology decision-maker within 14 hours of acute admission. There was no system in place for 'flagging' patients who had been admitted.	N	There was no service organisation policy.

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
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).	N	Only inpatients were discussed at the weekly haematology MDT meetings. Arrangements for discussions of patients from local teams or with complex issues were not held on a regular basis.	N	Multi-disciplinary team meetings to discuss and review patient care were not yet in place. Some patients would be discussed during the wider haematology MDT mtgs
HN-603	**Delegation of Annual Reviews** If annual reviews are undertaken by LHT/s on behalf of the SHT, a written agreement should be in place covering: a. Monitoring protocols (QS HN-502) b. LHT management and referral guidelines (QS HN-503) c. National Haemoglobinopathy Registry data collection (QS HN-701)	N	Arrangements were not yet in place. A clinic attended by the team from GSTT had not taken place for over 12 months	N	Arrangements were not yet in place
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		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		N/A	

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
HN-606	**Trans-Cranial Doppler Ultrasound Standard Operating Procedure (Children's Services Only)** A Standard Operating Procedure for Trans-Cranial Doppler ultrasound should be in use covering at least: a. Trans-Cranial Doppler modality used b. Identification of ultrasound equipment and maintenance arrangements c. Identification of staff performing Trans-Cranial Doppler ultrasound (QS HN-207) d. Arrangements for ensuring staff performing Trans-Cranial Doppler ultrasound have and maintain competences for this procedure, including action to be taken if a member of staff performs less than 40 scans per year e. Arrangements for recording and storing images and ensuring availability of images for subsequent review f. Reporting format g. Arrangements for documentation and communication of results h. Internal systems to assure quality, accuracy and verification of results	N/A		N	The guidance was due for review in 2017
HN-607	**Network Review and Learning Meetings** At least one representative of the team should attend each Network Review and Learning Meeting (QS HY-798).	N	There was no established network in place. Members of the team did attend UK Forum meetings and meetings with the Oxford Network	N	There was no established network in place.

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
HN-608	**Neonatal Screening Programme Review Meetings** The SHT should meet at least annually with representatives of the neonatal screening programme to review progress, discuss audit results (QS HN-703), identify issues of mutual concern and agree action.	Y		Y	
HN-701	**National Haemoglobinopathy Registry** Data on all patients, following patient or parental consent, should be entered into the National Haemoglobinopathy Registry. Data should include annual updates, serious adverse events, pregnancies and patients lost to follow up.	N	Patients who had consented, were registered on the NHR database by the lead nurse. Data on annual updates, serious adverse incidents were not yet entered.	N	Patients were registered on the NHR database by the lead nurse but data on annual updates, serious adverse incidents etc were not yet entered.
HN-702	**Activity Data** The service should monitor on an annual basis: a. Number of acute admissions, day unit admissions, Emergency Department attendances and out-patient attendances b. Length of in-patient stays c. Re-admission rate d. 'Did not attend' rate for out-patient appointments	Y		Y	

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
HN-703	**Quality Dashboard** The service should monitor Quality Dashboard data including the number and percentage of: a. Adverse events reported on the NHR for which a mortality or serious case review has taken place b. Children who have had Trans-Cranial Doppler screening undertaken within national guidelines c. Patients given pain relief within half an hour of presentation with sickle crisis d. Patients with possible sickle disorders identified by neonatal screening who have been entered on a care pathway e. Eligible children beginning penicillin at or before three months of age f. Patients registered on the National Haemoglobinopathy Registry who had an annual review undertaken within the last year g. Patients on long-term transfusion who received cardiac MRI, and the proportion of those receiving a cardiac MRI who achieved a figure of less than 20ms h. Eligible patients with sickle cell disease who received an MRI for liver iron, and the proportion of those who received an MRI for liver iron who achieved more than 7 mg/gm/DW (sickle cell and thalassaemia separately)	N/A	The adult service had received confirmation of Specialist Haemoglobinopathy Team (SHT) status as part of the national reorganisation of haemoglobinopathy services, so were planning to submit data to the quality dashboard in the future.	N	Evidence was not available to show compliance with this QS .
HN-704	**Other Quality Data** The service should monitor on an annual basis: a. Proportion of children who have been offered treatment following Trans-Cranial Doppler Screening	N/A		Y	

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	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)	N	A records audit had been undertaken but no other audits as required by the QS. No admin or clerical support was available to help with data management.	N	A records audit had been undertaken but no other audits as required by the QS. No admin or clerical support was available to help with data management.
HN-706	**Network Audits** The service should participate in agreed network-wide audits.	N	A network audit programme was not in place.	N	A network audit programme was not in place.
HN-707	**Research** The SHT should actively participate in research relating to the care of patients with haemoglobin disorders.	N	The service was not yet involved in, or referred patients for inclusion in research taking place elsewhere relating to the care of patients with haemoglobin disorders.	N	The service was not yet involved in, or referred patients for inclusion in research taking place elsewhere relating to the care of children and young people with haemoglobin disorders.

		Adults- SHT		Children - SHT	
Ref	Standard	Met?	Comments	Met?	Comments
HN-708	**Trans-Cranial Doppler Quality Assurance (Children's Services Only)** The service should monitor and review at least annually: a. Whether all staff performing Trans-Cranial Doppler ultrasound have undertaken 40 procedures in the last year (QS HN-207) b. Results of internal quality assurance systems (QS HN-606) c. Results of National Quality Assurance Scheme (NQAS) for Trans-Cranial Doppler Ultrasound (when established) or local peer review arrangements (until NQAS established)	N/A		N	Evidence that internal quality assurance had taken place was not available. 'c ' was not applicable as a national process had not yet been formalised.
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)	N	Data were not collected for 'c' to enable multidisciplinary review.	N	Data were not collected for 'c' to enable multidisciplinary review.
HN-799	**Document Control** All policies, procedures and guidelines should comply with Trust (or equivalent) document control procedures.	N	See main report in relation to out of date guidance.	N	See main report in relation to out of date guidance.

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