MLCSU Clinical

NHS
Midlands and Lancashire
Commissioning Support Unit

UK FORUM FOR HAEMOGLOBIN DISORDERS

Health Services for People with Haemoglobin Disorders

Alder Hey Children's NHS Foundation Trust

Visit date: 28th February 2024

Report date: 11th July 2024

Contents

Introduction

This report presents the findings of the review of Alder Hey Children's NHS Foundation Trust that took place on 28th February 2024. The purpose of the visit was to review compliance with the Health Services for People with Haemoglobin Disorders Quality Standards Version 5.1, November 2021 which were developed by the UK Forum for Haemoglobin Disorders (UKFHD). The peer review programme and visit was organised by the Nursing and Urgent Care Team (NUCT) at the Midlands and Lancashire Commissioning Support Unit (MLCSU). The Quality Standards refer to the following types of specialised service for people with haemoglobin disorders:

- Specialist Haemoglobinopathy Team
- Local Haemoglobinopathy Team (or Linked Provider)

A comprehensive peer review of Local Haemoglobinopathy Teams (LHT) against the Local Haemoglobinopathy Team Quality Standards was not part of the 2024 -2026 programme, however Haemoglobinopathy Coordinating Centres were given the option to request a review visit for any of their Local Haemoglobinopathy Teams in their review visit programme.

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 risks identified will include the Trust and UKFHD/MLSCU response to any actions taken to mitigate against the risk. Appendix 1 lists the visiting team that reviewed the services in Alder Hey health economy. Appendix 2 contains the details of compliance with each of the standards and the percentage of standards met.

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

- Alder Hey Children's NHS Foundation Trust
- NHS England – North West
- NHS Cheshire and Merseyside Integrated Care Board

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 commissioners in relation to this report is NHS Cheshire and Mersey Integrated Care Board.

About the UKFHD and MLCSU

The UK Forum on Haemoglobin Disorders (UKFHD) is a multi-disciplinary group of healthcare professionals interested in all aspects of sickle cell disease, thalassaemia, and related conditions. The Forum is now a recognised and respected organisation involved in formulating national policy for screening and management of these conditions. The UKFHD aims to ensure equal access to optimal care for all individuals living with an inherited haemoglobin disorder or rare inherited anaemia. The mission of the UKFHD is to, advocate and influence policy, promote and review best practice, share ideas, and advise on research, educate health professionals, and support education of patients, whilst influencing and advocating on equitable prevention programmes for sickle cell and thalassaemia disorders.

The Midlands and Lancashire Commissioning Support Unit (MLCSU) Nursing and Urgent Care Team (NUCT) is a trusted partner for specialist, independent, clinical, and analytical guidance on a regional, national, and international scale. Our team has significant experience in developing, facilitating, and delivering peer review programmes.

More detail about the work of the UKFHD and the MLCSU is available at https://haemoglobin.org.uk/ and https://www.midlandsandlancashirecsu.nhs.uk/our-expertise/nursing-and-urgent-care/

Acknowledgements

The UKFHD and MLCSU would like to thank the staff and service users and carers of the Alder Hey Health economy for their hard work in preparing for the review and for their kindness and helpfulness during the course of the visit. Thanks, are also due to the visiting team and their employing organisations for the time and expertise they contributed to this review.

Return to Index

Review Visit Findings

Trust-wide General Comments

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

Alder Hey Hospital provided acute and tertiary specialist services for children residing across the North West, North Wales, Isle of Man and parts Cheshire.

The Trust was a provider of specialist services for haemoglobin disorders[1] and had been formally recognised as a joint Specialist Haemoglobinopathy Team with Liverpool University Hospital Foundation Trust in April 2019 following the development of a new national service specification framework by NHS England (NHSE).

Some issues in this report relate specifically to the Trust as a whole and have been included in the Trust-wide section of the report. Other issues that are the same for the children and young people service have been repeated in each section. Further support from the Trust executive team and commissioners will also be required for the paediatric SHT to fulfil its role.

Trust-wide Good Practice

1. The staff competence framework was very good with clearly defined objectives for all levels of staff. The framework had been adapted from the RCN Nurse competencies and was available on the staff training portal. All nurses regularly caring for children with a haemoglobinopathy condition had to complete the competencies and be reassessed every year. Band 5 and 6 nurses were also given the opportunity to shadow the Band 7 CNS to gain more experience. The CNSs also had plans to implement a similar process for staff working the emergency department and other admitting areas.
2. Reviewers were impressed with the work of the Psychologist, who with limited time (0.4wte), was working above and beyond her role in terms of the support given to children and their families. Parents who met the visiting team were extremely grateful for the holistic approach and support they had received.

Trust-Wide Concern

1. Consultant staffing

Reviewers were concerned that the service had insufficient consultant medical staff with appropriate competences in the care of people with haemoglobin disorders. Out of an establishment of five WTE consultant haematologists only four substantive haematologists were in post and there were gaps in the day-to-day provision due to long term sickness of one consultant and a recurrent failure to recruit to the vacant post.

To mitigate the issue the Trust had implemented a structured model whereby a locum consultant covered all paediatric haematology outpatient and day unit care, and the other three consultants covered the on call and ward rotas.

The management team who met with the visiting team were monitoring the situation and the shortage of consultant staff was documented on the Trust risk register.

2. Access to psychology

[1] As a partnership SHT with Liverpool University Hospital Foundation Trust the term paediatric SHT has been used throughout the report to refer to the services specifically provided by Alder Hey Children's NHSFT

The service had 0.4wte psychologist for 153 patients which just met the British Psychological Society Special Interest Group in Sickle Cell and Thalassaemia (2017) recommendation of one WTE psychologist for every 300 patients but would be insufficient time as activity increases, which is likely year on year. At the time of visit the postholder was on maternity leave and there was limited access to psychology support in their absence. Reviewers were told that urgent referrals would be seen by the trust service but there was a significant wait for routine referrals including neurocognitive assessments.

3. Access to welfare and benefits support

Patients with haemoglobinopathy disorders did not have easy access to social, immigration or benefits advice which resulted in clinical staff spending a considerable amount of time supporting patients and their families with welfare and immigration issues. Some support would be available to patients via the Roald Dahl Foundation, but the foundation would not be able to provide a comprehensive service.

Trust-wide Further Considerations

1. Training for staff in the ED and for those working on non-designated haematology wards should be reviewed especially following feedback from service user and families who reported frustration about the lack of knowledge staff had about their child's condition, and that patients were not always admitted on haematology wards.
2. The trust did not have a formal service level agreement covering the SHT joint working and financial arrangements with Liverpool University Foundation Trust. Reviewers considered that it would be sensible for both trusts to clarify the responsibility for each centre and the finances associated with the partnership.
3. Reviewers did not meet any commissioners from NHSE or the local integrated care board during the visit and due to changes in the NHSE regional team the paediatric SHT were not clear who to contact. As commissioning arrangements change over the next few years it will be important to build these relationships.
4. Some of the patients who met with the reviewing team spoke about the stigma of being labelled by their condition in the ED and other outlying areas. The Trust communication team should reiterate to staff the importance of being mindful of how people are addressed.

Views of Service Users and Carers

Support Group available for patients and carers	Y/N
Sickle Cell Disease – Children and Young People	Y
Thalassaemia – Children and Young People	N

During the visit the visiting team met with three parents and two young people living with sickle cell disease and one parent with the help of an interpreter) who had a child living with thalassaemia.

The views of the users were wide-ranging and are documented in the children's and young people specialist haemoglobinopathy team section. The review team would like to thank them for their openness and willingness to share their experiences.

Return to Index

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

General Comments and Achievements

This was an experienced service with strong leadership evident throughout. The team were extremely proud of what they had achieved since the last visit in 2019 and it was clear to the reviewers that the team was highly committed and enthusiastic. The Haemoglobinopathy Team worked hard to ensure children's conditions were managed well within the community to prevent admissions to hospital and to ensure patients received a high standard of care within the inpatient setting.

There had been an increase in activity since the review in 2019 from 75 to 153 patients with a haemoglobin disorder. The paediatric SHT had 10 local haemoglobinopathy teams (LHTs), within their catchment, all were low prevalence areas.

The Haemoglobinopathy Team consisted of four Consultant Haematologists and a locum Consultant who had joined the Team in May 2023. There were two Clinical Nurse Specialists (1.7wte) and some support from a Haematology Associate CNS. The team had 0.4wte Clinical Psychology support and a Specialist Haemoglobinopathies Midwife in each maternity unit.

Local haemoglobinopathy teams could access the paediatric SHT for advice during normal working hours and at weekends via the non-resident doctor on call. Patients would have their bloods tests locally at their LHT and then all patients were followed up a telephone call by the SHT CNS. LHTs representatives who spoke to the reviewing teams said access to advice was straight forward with clear plans either by email or phone if time critical and good support from the paediatric SHT at other times. Parents liked the specialist care they received at Alder Hey but also the access to local care closer to home. There were good relationships between the local community nurses and paediatric SHT CNS team.

The Trust had a 24/7 contract to provide red cell exchange with NHSBT and surgical vascular access was always available.

All annual reviews and transcranial doppler scans (TCD) were undertaken by the paediatric SHT at Alder Hey. TCDs were performed by two practitioners one of whom was the joint lead for TCD for the North West Sickle Cell HCC.

An efficient nurse-led hydroxycarbamide service was in operation.

Transition commenced from the age of 13 years with self-assessment questionnaires and a workbook to educate the young adult and prepare them for independence as an adult. Transfer to adult services occurred from the age of 16 years, depending on the young person's wishes and readiness for transition.

<table>
<tr><th colspan="4">SPECIALIST HAEMOGLOBINOPATHY TEAM - CHILDREN AND YOUNG PEOPLE</th></tr>
<tr><td rowspan="4">Alder Hey Children's Hospital</td><td colspan="3">Linked Haemoglobinopathy Coordinating Centres (HCC)</td></tr>
<tr><td colspan="3">North West Sickle Cell HCC
North of England Thalassaemia and RIA HCC</td></tr>
<tr><td rowspan="2">Linked Local Haemoglobinopathy Teams (LHT)</td><td colspan="2">Patient Distribution</td></tr>
<tr><td>SCD</td><td>Thal.</td></tr>
<tr><td>Betsi Cadwaladr University Health Board - Bangor Hospital, Glan Clwyd Hospital, Wrexham Maelor Hospital</td><td>12</td><td>1</td></tr>
</table>

	Countess of Chester NHS Trust	1	1
	Mid Cheshire Hospitals NHS Foundation Trust	2	0
	Noble's Hospital, Isle of Man	2	0
	St Helens and Knowsley Teaching Hospitals NHS Trust	6	0
	Southport and Ormskirk Hospital NHS Trust	1	4
	Warrington and Halton Hospitals NHS Foundation Trust	2	1
	Wirral University Teaching Hospital NHS Foundation Trust	5	1

PATIENTS USUALLY SEEN BY THE CHILDREN AND YOUNG PEOPLE SPECIALIST HAEMOGLOBINOPATHY TEAM						
Condition	Registered patients	Active patients *[2]	Annual review **	Long-term transfusion	Eligible patients - hydroxycarbamide	In-patient admissions in last year
Sickle Cell Disease	120	120	120	7	84/86	31 admissions 2022-23 totalling 171 inpatient days 47 admissions totalling 147 days Apr 2023 -Jan 2024
Thalassaemia	33	33	33	7	N/A	0 admissions for 2022-23 1 admission totalling 5 days in April 2023 – Jan 24

Staffing

Specialist Haemoglobinopathy Team	Number of patients	Actual WTE (at time of visit)
Consultant haematologist/paediatrician dedicated to work with patients with hemoglobinopathies	153	4 WTE Consultant Haematologists with a total of 4.5PAs time for HBO
Clinical Nurse Specialist for paediatric patients dedicated to work with patients with hemoglobinopathies	153	2 CNS total of 1.8 WTE
Clinical Psychologist for paediatric patients dedicated to work with patients with hemoglobinopathies	153	0.4 WTE psychologist

[2] ** Those who have had hospital contact in the last 12 months. **No. patients who have had an annual review in last year*

Urgent and Emergency Care

All children and young people needing an acute assessment or admission were seen via the children's Emergency Department (ED) where they would receive a medical review depending on their triage rating. The team were providing patients with an ED blue card which would indicate Priority Triage when they attended. National Haemoglobinopathy Patient Cards were also in use. Patients had individualised emergency care plans which were part of their annual review, and these were filed in their medical records and accessible to ED staff. Guidelines for professionals to access were held on the Trust intranet document management system. Staff in the ED could contact the on-call Consultant Haematologist at any time (both in and out of working hours), within working hours they called the Haematology Registrar or Clinical Fellow and Nurse Specialists to seek advice. The paediatric SHT had delivered bitesize training on haemoglobinopathy conditions and complications to ED staff at their lunch time sessions and a clinical simulation related to sickle cell patients, usually involving acute chest syndrome was held annually between ED and the haemoglobinopathy team.

In-patient Care

The new in-patient ward was co-located with the outpatient clinic and day care facility on Ward 3b and the main ward for all haemoglobinopathy patients. The Ward was a 13 bedded dedicated haematology/oncology unit which consisted of 6 en-suite cubicles and 2 transplant rooms. In addition, the ward had a 4 bedded bay area (only one bed used at a time due to IPC regulations) as well as 4 en-suite cubicles in the Teenage Cancer Trust Unit (TCT). The TCT had a chill out room, plus an activity area, and teenagers with Haemoglobinopathies have access to this room. The ward had a school room with dedicated teacher and Play Specialists to provide therapeutic play activities for children of all ages and abilities. The ward also had a dedicated kitchen with chefs to cook to order. A parent/carer bed was situated next to every child's bed. Accommodation was also available, where appropriate, in Ronald McDonald House. Outlying patients were admitted to Ward 3c (Medical Specialty Ward) whenever possible and occasionally patients were admitted to other wards within the hospital and were assessed on their priority for transfer to Ward 3b.

Medical ward rounds took place daily by either a haematology or oncology consultant. All children with haemoglobin disorders were admitted under the care of the attending haematology consultant.

Day and outpatient care

The day care facility on ward 3b was open between 0800 to 1845 for planned admissions, and between 0800 to 16.30 for unplanned admissions and used by haemoglobinopathy patients for transfusions, treatment, and clinical reviews. The facility had 10 commissioned beds: four bedded bay, two cubicles, a two bedded bay for teenagers and young adults, and two treatment chairs.

Nurses on the day care facility were trained in cannulation and accessing Central Venous Access Devices.

Outpatient clinics were held in the consulting rooms within 3B. There was a waiting area with play facilities, a height/weight room, and a quiet room. A dedicated sickle cell clinic was held on a Thursday afternoon every week and a weekly nurse led hydroxycarbamide monitoring clinic Wednesday afternoon with ad-hoc clinics on a Thursday afternoon when necessary.

Teenage/Transition clinics were held in conjunction with the adult team four times a year at Alder Hey and a young person's clinic was held twice a year with the adult SHT at the Royal Liverpool University Hospital.

A range of joint speciality clinics were in operation. A combined sickle cell/respiratory clinic was held every two months; a nephrology MDT was held twice a year to discuss patient referrals and eligibility for review via he the nephrology clinic and a neurology clinic was held four times per year as well as a joint MDT twice a year.

Phlebotomy was provided in the outpatient and daycare departments Monday-Thursday 9am-4.30pm and Friday 9am-12 noon.

Community-based Care

Both Clinical Nurse Specialists offered community-based care which included the giving of newborn screening results, school visits, home visits and liaison with other professionals.

Views of Service Users and Carers

The visiting team met with three parents and two young people living with sickle cell disease and one parent with the help of an interpreter) who had a child living with thalassaemia during the visit. See their feedback below about the services provided by the Trust.

Service carer feedback relating to Thalassaemia care and support.

- The parent was happy with the treatment and care they had received and had not needed to access any emergency treatment. They were aware that they could call the CNS for advice on weekdays, though with language barrier they seemed unclear about the process for accessing advice over the weekends. The CNS's had visited the child's school and had explained about iron chelation.
- The child was being considered for transplant but had not been offered any psychology support and the parents understanding via the interpreter was that their child had to be 'transfused until cured'.

Service user and carers feedback relating to Sickle Cell care and support.

- The children who met with the visiting team talked about the good education around their condition to the extent that they thought they could present to others about sickle cell disease.
- The Paediatric SHT team were wonderful.
- The CNSs had attended their children's schools to provide education for staff and reiterate the importance of their children having adequate fluid during the day and the need to be excused for regular toilet breaks.
- One of the parents had been so inspired by the team they were now training to be a nurse.
- Psychology support was good and the implementation of annual neuro cognitive 'assessment had provided reassurance. Others spoke about the support from the psychologist who had helped them with the guilt they felt when finding out of the diagnosis when they were pregnant.
- Pre-natal and post-natal counselling had been very good with the introduction to the team at Alder Hey. One parent talked about the support they had received when their baby was newly diagnosed, commenting that the first month following diagnosis was the hardest.
- One parent had a 16yr old and twin 14-year-olds. The eldest child was in the process of transiting to adult care and was not keen to move to the adult service which was causing some angst with the younger children who had not yet commented on their transition pathway.
- The child of one parent had been critically ill and had required an intensive care admission. The parent was clearly very traumatised by the whole episode and still felt that there had not been enough urgency and understanding by staff in ED. The child had been seen within 25 minutes, but administration of analgesia was delayed. In both the ED and ITU areas they believed staff thought it was their fault that their child had become so critically unwell. The paediatric SHT had been fantastic in the level of support they had given them.
- Others commented that they could not fault the care in the ED but also commented that had heard staff call their child the 'a sickle cell child' before being called in for an assessment and the stigma they felt this caused.
- Comments were made that their child's GPs were reluctant to initiate treatments and tended to send them to hospital rather than deal with their child's complex or acute pain.

- When asked about the care on the ward one parent commented that their child had never been admitted the haematology ward.
- They spoke highly of community CNS role and were very concerned about the imminent retirement of one of the CNSs' and whether they would receive the same level of care and support in the future.

Good Practice

1. The work of the CNS team and their focus and commitment to providing holistic care for children, young people, and their families living with a haemoglobinopathy condition was commendable. Reviewers were particularly impressed with the following:

    a. The outreach model provided good continuity of care and was highly valued by the parents and children who met with the visiting team. The CNS were proactive in schools to ensure that education staff were aware of the child's care plan and to provide education. Patients could have they bloods samples taken in the community setting which reduced the number of times they had to attend the hospital.

    b. Those families with a newly affected infant were seen by the CNS at the family home within 5 days of the screening result. The CNS would also undertake a follow up visit the following day which reviewers considered was a very good way ensuring that the family was given some time to understand the implications of the results, and for the CNS to provide advice and support. Parents who met with the visiting team commented on the value of having this level of support at the difficult time of their child's diagnosis.

    c. The CNSs had initiated the idea of an education board game for children with sickle cell disease and then worked collaboratively with Roald Dhal Foundation to develop this into a commercial product. The board game provided a fun way to give children information and assess their understanding of their condition. The CNS were also hoping that a similar product could be developed for children with thalassaemia.

    d. The level of teaching provided was comprehensive and it was delightful to hear from two young adults at the visit that they felt confident and knowledgeable enough to provide teaching to others about their condition.

2. The staff competence framework was very good with clearly defined objectives for all levels of staff. The framework had been adapted from the RCN Nurse competencies and was available on the staff training portal. All nurses regularly caring for children with a haemoglobinopathy condition had to complete the competencies and be reassessed every year. Band 5 and 6 nurses were also given the opportunity to shadow the Band 7 CNS to gain more experience. The CNSs also had plans to implement a similar process for staff working the emergency department and other admitting areas.

3. Reviewers were impressed with the work of the Psychologist who, with limited time (0.4wte), was working above and beyond her role in terms of the support given to children and their families. Parents who met the visiting team were extremely grateful for the holistic approach and support they had received.

    The psychologist would also visit schools to support children and particularly to explain to education staff the rationale for providing reasonable adjustments for children to promote their health and wellbeing whilst at school.

    The psychologist had also developed a neuro cognitive screening assessment tool which was undertaken on annual basis at the time of the child's annual review. Parents commented that use of the tool annually provided then with assurance that any cognitive issues would be identified quickly.

4. The screening fail safe mechanism was well thought through with a robust audit process in place. The laboratory would send a monthly list of carrier results to the screening link health visitor who would visit

the family at home to give the results. If any further counselling was required, then the health visitor would refer to the CNS team. Affected infant's results were sent to the CNS team who would visit the parents at home within 5 working days of receiving the results from the screening laboratory. For both processes an acknowledgement that the information had been received was required and documented.

A haemoglobinopathy screening meeting was held twice a year with all professionals involved in antenatal and newborn screening to review the screening and audit processes as well as providing up to date information on any increasing activity.

5. The review team met with one of the LHTs who valued the three-monthly update meetings provided by the paediatric SHT which was a useful opportunity for the paediatric SHT to check any changes in the number of patients attending LHT.
6. The paediatric SHT had an impressive and active audit programme. Audits were well analysed, with documented outcomes, discussion points and actions identified which were then included in the paediatric SHT annual report.

Immediate Risk

The review team identified no immediate risks during the visit.

Concerns

1. Consultant staffing

Reviewers were concerned that the service had insufficient consultant medical staff with appropriate competences in the care of people with haemoglobin disorders. Out of an establishment of five WTE consultant haematologists only four substantive haematologists were in post and there were gaps in the day-to-day provision due to long term sickness of one consultant and a recurrent inability to recruit to the vacant post.

To mitigate the issue the Trust had implemented a structured model whereby a locum consultant covered all paediatric haematology outpatient and day unit care, and the other three consultants covered the on call and ward rotas.

The management team who met with the visiting team were monitoring the situation and the shortage of consultant staff was documented on the Trust risk register.

2. Access to psychology

The service had 0.4wte psychologist for 153 patients which just met the British Psychological Society Special Interest Group in Sickle Cell and Thalassaemia (2017) recommendation of one wte psychologist for every 300 patients but would be insufficient time as activity increases which is likely year on year. At the time of visit the postholder was on maternity leave and there was limited access to psychology support in their absence. Reviewers were told that urgent referrals would be seen by the trust service but there was a significant wait for routine referrals including neurocognitive assessments.

3. Access to welfare and benefits support

Patients with hemoglobinopathy disorders did not have easy access to social, immigration or benefits advice which resulted in clinical staff spending a considerable amount of time supporting patients and their families with welfare and immigration issues. Some support would be available to patients via the Roald Dahl Foundation, but the foundation would not be able to provide a comprehensive service.

Further Considerations

1. The paediatric SHT was about to go through a significant change to its CNS establishment and it will be important to maintain patient confidence, particularly as patients who met with the reviewing team were very concerned about the future of the community service.

Senior management and the paediatric SHT were undertaking a skill mix and service review to identify solutions to maintain service delivery due to the imminent retirement of a very experienced member of the team and to future proof the service. Reviewers were supportive of the plan for the existing postholder to provide a mentoring function as it was unlikely they would be able to recruit a specialist nurse with experience in haemoglobinopathy disorders. Reviewers were also made aware that a band 8a post across haematology was also being considered, although it was not clear how much of this role would be providing senior support to the paediatric SHT and whether this role would help address the increasing activity and workload being experienced by the paediatric SHT.

2. The paediatric SHT had undertaken an audit of compliance with NICE Clinical Guideline on the management of acute pain but due to the small numbers in the audit, changing to a prospective audit may a be more useful methodology to adopt.
3. Training for staff in the ED and for those working on non-designated haematology wards should be reviewed especially following feedback from service user and families who reported frustration about the lack of knowledge staff had about their child's condition and that patients were not always admitted to haematology wards.
4. The paediatric SHT team were regular attenders at the HCC MDT and presented cases. They felt that they would benefit from participation in MDT discussion of cases which reflected the full geographical area of the HCC. This has also been reflected in the HCC report.
5. The trust did not have a formal service level agreement covering the paediatric SHT joint working and financial arrangements with Liverpool University Foundation Trust. Reviewers considered that it would be sensible for both trusts to clarify the responsibility for each centre and the finances associated with the partnership.
6. Reviewers did not meet any commissioners from NHSE or the local integrated care board during the visit and due to changes in the NHSE regional team the paediatric SHT were not clear who to contact. As commissioning arrangements change over the next few years it will be important to build these relationships.
7. Some of the patients who met with the reviewing team spoke about the stigma of being labelled by their condition in the ED and other outlying areas. Trust communication team should reiterate to staff the importance of being mindful of how people are addressed.
8. The patient support group had recommenced in November 2023 and patient feedback was that support networks were important for providing peer support with other parents who were often isolated from within their local community.

Commissioning

The review team did not meet with any NHS England or local integrated care board commissioners. Several of the issues in this report will require the active involvement of the Trust leadership team and commissioners to ensure that timely progress is made.

Return to Index

Appendix 1 - Membership of Visiting Team

Visiting Team		
Clare Clark	Paediatric Haematology Clinical Nurse Specialist	Cambridge University Hospital
Leah Denver	Network Manager	Birmingham Women & Children's Hospital NHS Trust
Rosie Peregrine-Jones	Assistant Director of Quality	North Central London ICB
Romaine Maharaj	Chief Executive	UK Thalassemia Society

Clinical Lead		
Mark Velangi	Consultant Paediatric Haematologist	Birmingham Women & Children's Hospital NHS Trust

MLCSU Team		
Sarah Broomhead	Professional Lead	MLCSU
Gill Hay	PA Nursing and Urgent Care	MLCSU

Return to Index

Appendix 2 - Compliance with the Quality Standards

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

Percentage of Quality Standards met

Service	Number of Applicable QS	Number of QS Met	% Met
Specialist Haemoglobinopathy Team (SHT) Children and Young People	49	43	90%

Specialist Haemoglobinopathy Team for Children and Young People with Haemoglobin Disorders

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-101	**Haemoglobin Disorder Service Information** Written information should be offered to children, young people and their families, and should be easily available within patient areas, 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 Contact interpreter and advocacy services, Patient Advice and Liaison Service (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 HC-199)	Y	The team were in the process of implementing access to information via QS codes.

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-102	**Information about Haemoglobin Disorders** Children, young people and their families should be offered written information, or written guidance on where to access information, covering at least: a) A description of their condition (SCD or Th), how it might affect them and treatment available b) Inheritance of the condition and implications for fertility c) Problems, symptoms and signs for which emergency advice should be sought d) How to manage pain at home (SCD only) e) Transfusion and iron chelation f) Possible complications g) Health promotion, including: i. Travel advice ii. Vaccination advice h) National Haemoglobinopathy Registry, its purpose and benefits i) Parental or self-administration of medications and infusions	Y	
HC-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. Planned acute and long-term management of their condition, including medication iii. 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 patient's GP and their local team consultant (if applicable).	Y	Clinic letters were seen for those patients with SCD, who were non transfusion dependant and transfusion dependant. All letters included contact details and condition specific information.
HC-104	**What to Do in an Emergency?** All children and young people should be offered information about what to do in an emergency covering at least: a. Where to go in an emergency b. Pain relief and usual baseline oxygen level, if abnormal (SCD only)	Y	

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-105	**Information for Primary Health Care Team** Written information, or written guidance on where to access 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 (SCD and splenectomised Th) and analgesia (SCD) b. Side effects of medication, including chelator agents [SCD and Th] c. Guidance for GPs on: i. Immunisations ii. Contraception and sexual health (if appropriate) d. What to do in an emergency e. Indications and arrangements for seeking advice from the specialist service	Y	
HC-106	**Information about Transcranial Doppler Ultrasound** Written information should be offered to children, young people and their families covering: a. Reason for the scan and information about the procedure b. Details of where and when the scan will take place and how to change an appointment c. Any side effects d. Informing staff if the child is unwell or has been unwell in the last week e. How, when and by whom results will be communicated	Y	
HC-107	**School or College Care Plan** A School or College Care Plan should be agreed for each child or young person covering at least: a. School or college attended b. Medication, including arrangements for giving / supervising medication by school or college staff c. What to do in an emergency whilst in school or college d. Arrangements for liaison with the school or college e. Specific health or education need (if any)	Y	

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-194	**Environment and Facilities** The environment and facilities in phlebotomy, outpatient 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 age-appropriate toys, reading materials and multimedia. There should be sound and visual separation from adult patients.	Y	
HC-195	**Transition to Adult Services** Young people approaching the time when their care will transfer to adult services should be offered: a. Information and support on taking responsibility for their own care b. The opportunity to discuss the transfer of care at a joint meeting with paediatric and adult services c. A named coordinator for the transfer of care d. A preparation period prior to transfer e. Written information about the transfer of care including arrangements for monitoring during the time immediately after transfer to adult care f. Advice for young people leaving home or studying away from home including: i. Registering with a GP ii. How to access emergency and routine care iii. How to access support from their specialist service iv. Communication with their new GP	Y	A good SOP was in place covering transition and information leaflets for young people detailing the process
HC-197	**Gathering Views of Children, Young People and their Families** The service should gather the views of children, young people and their families at least every three years using: a. 'Children's Survey for Children with Sickle Cell' and 'Parents Survey for Parents with Sickle Cell Disorder' b. UKTS Survey for Parents of Children with Thalassaemia	Y	SCD Picker survey had been undertaken in 2023. A Thalassaemia and RIA survey had commenced December 2023 Responses to the surveys had been low so the team were planning to repeat later in the year.

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-199	**Involving Children, Young People and Families** The service's involvement of children, young people and their families should include: a. Mechanisms for receiving feedback b. Mechanisms for involving children, young people and their families in: i. Decisions about the organisation of the service ii. Discussion of patient experience and clinical outcomes (QS HC-797) c. Examples of changes made as a result of feedback and involvement	Y	Following feedback, the team had changed the day of the week clinics were held.
HC-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. The lead consultant should undertake Continuing Professional Development (CPD) of relevance to this role, should have an appropriate number of session/s identified for the role within their job plan and cover for absences should be available.	Y	The peadiatric lead/deputy for SHT had 1 PA for network duties. There was a named deputy, but the role had not yet been formalised.
HC-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 children and young people with haemoglobin disorders The lead nurse should have appropriate time for their leadership role and cover for absences should be available.	Y	2 CNS who shared the role (0.9wte and 0.8wte)
HC-204	**Medical Staffing and Competences: Clinics and Regular Reviews** The service should have sufficient medical staff with appropriate competences in the care of children and young people with haemoglobin disorders for clinics and regular reviews. Competences should be maintained through appropriate CPD. Staffing levels should be appropriate for the number of patients cared for by the service and its role. Cover for absences should be available.	N	4.5 PAs dedicated for haemoglobinopathy work for the 153 patients registered with the paediatric SHT.

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-205	**Medical Staffing and Competences: Unscheduled Care** 24/7 consultant and junior staffing for unscheduled care should be available. **SHTs and HCCs only:** A consultant specialising in the care of children and young people with haemoglobin disorders should be on call and available to see patients during normal working hours. Cover for absences should be available.	Y	
HC-206	**Doctors in Training** If doctors in training are part of achieving QSs HC-204 or HC-205 then they should have the opportunity to gain competences in all aspects of the care of children and young people with haemoglobin disorders.	Y	
HC-207	**Nurse Staffing and Competences** The service should have sufficient nursing staff with appropriate competences in the care of children and young 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 or other staff 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. Cover for absences should be available.	N	The two Clinical Nurse Specialists (1.7wte) covered both the acute and community Service. One CNS (0.8 wte) was retiring at the end of May.

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-208	**Psychology Staffing and Competences** The service should have sufficient psychology staff with appropriate competences in the care of children and young 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 multidisciplinary discussions and governance activities c. Provision of, or arrangements for liaison with and referral to, neuropsychology Staffing levels should be appropriate for the number of patients cared for by the service and its role. Cover for absences should be available.	N	At the time of the visit there was 0.4wte psychology staffing for 153 patients, which did not meet the recommended staffing level of 1wte :300 patients. At the time of the visit the post holder was on maternity leave. Cover in the interim was mainly for urgent referrals and there was a long waiting time for routine referrals to be seen.
HC-209	**Transcranial Doppler Ultrasound Competences** Sufficient staff with appropriate competences for Transcranial Doppler ultrasound should be available. Staff should undertake at least 40 scans per annum and complete an annual assessment of competence. Cover for absences should be available.	Y	
HC-299	**Administrative, Clerical and Data Collection Support** Administrative, clerical and data collection support should be appropriate for the number of patients cared for by the service.	Y	
HC-301	**Support Services** Timely access to the following services should be available with sufficient time for patient care and attending multidisciplinary meetings (QS HC-602) as required: a. Social worker / benefits adviser b. Play specialist / youth worker c. Dietetics d. Physiotherapy (inpatient and community-based) e. Occupational therapy f. Child and adolescent mental health services	Y	

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-302	**Specialist Support** Access to the following specialist staff and services should be easily available: a. DNA studies b. Genetic counselling c. Sleep studies d. Diagnostic radiology e. Manual exchange transfusion (24/7) f. Automated red cell exchange transfusion (24/7) g. Pain team including specialist monitoring of patients with complex analgesia needs h. Level 2 and 3 critical care	Y	
HC-303	**Laboratory Services** UKAS / CPA accredited laboratory services with satisfactory performance in the NEQAS haemoglobinopathy scheme and MHRA compliance for transfusion should be available.	Y	
HC-304	**Urgent Care – Staff Competences** Medical and nursing staff working in the Emergency Departments and admission units should have competences in urgent care of children and young people with haemoglobin disorders.	N	Training was provided but a framework to assess competencies for staff working in urgent and emergency care areas was not yet in place. Simulation training was provided twice a week which had included scenarios in acute management of SCD

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-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 family or carer, paediatric and adult services, primary health care and social care in planning the transfer, including a joint meeting to plan the transfer of care 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 HCCs, SHTs and LHTs (if applicable) h. Responsibilities for giving information to the young person and their family or carer (QS HC-195)	Y	
HC-502	**New Patient and Annual Review Guidelines** Guidelines or templates should be in use covering: a. First outpatient appointment b. Annual review Guidelines should cover both clinical practice and information for children, young people and their families.	Y	

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-504	**Transcranial Doppler Ultrasound Standard Operating Procedure** A Standard Operating Procedure for Transcranial Doppler ultrasound should be in use covering at least: a. Transcranial Doppler modality used b. Identification of ultrasound equipment and maintenance arrangements c. Identification of staff performing Transcranial Doppler ultrasound (QS HC-209) d. Arrangements for ensuring staff performing Transcranial 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	Y	

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-505	**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: i. Manual exchange transfusion ii. Automated exchange transfusion on site or organised by another provider c. Investigations and vaccinations prior to first transfusion d. Recommended number of cannulation attempts e. Arrangements for accessing staff with cannulation competences f. Patient pathway and expected timescales for regular transfusions, including availability of out of hours services (where appropriate) and expected maximum waiting times for phlebotomy, cannulation and setting up the transfusion g. Patient pathway for Central Venous Access Device insertion, management and removal	Y	
HC-506	**Chelation Therapy** Guidelines on chelation therapy should be in use covering: Indications for chelation therapy a. Choice of chelation drug/s, dosage and dosage adjustment b. Monitoring of haemoglobin levels prior to transfusion c. Management and monitoring of iron overload, including management of chelator side effects d. Use of non-invasive estimation of organ-specific iron overloading heart and liver by T2*/R2 e. Self-administration of medications and infusions and encouraging patient and family involvement in monitoring wherever possible	Y	

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-507	**Hydroxycarbamide and Other Disease Modifying Therapies** Guidelines on hydroxycarbamide and other disease modifying therapies should be in use covering: a. Indications for initiation b. Monitoring of compliance and clinical response, including achieving maximum tolerated dose for hydroxycarbamide c. Documenting reasons for non-compliance d. Monitoring complications e. Indications for discontinuation	Y	
HC-508	**Non-Transfusion Dependent Thalassaemia (nTDT)** Guidelines on the management of Non-Transfusion Dependent Thalassaemia should be in use, covering: a. Indications for transfusion b. Monitoring iron loading c. Indications for splenectomy d. Consideration of options for disease modifying therapy	Y	
HC-509	**Clinical Guidelines: Acute Complications** Guidelines on the management of the acute complications listed below should be in use covering at least: i. Local management ii. Indications for seeking advice from the HCC / SHT iii. Indications for seeking advice from and referral to other services, including details of the service to which patients should be referred For children and young people with sickle cell disorder: 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 For children and young people with thalassaemia: l. Fever, infection and overwhelming sepsis m. Cardiac, hepatic or endocrine decompensation	Y	

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-510	**Clinical Guidelines: Chronic Complication** Guidelines on the management of the chronic complications listed below should be in use covering at least: i. Local management ii. Indications for discussion at the HCC MDT iii. Indications for seeking advice from and referral to other services, including details of the service to which patients should be referred iv. Arrangements for specialist multidisciplinary review a. Renal disease, including sickle nephropathy b. Orthopaedic problems, including the management of sickle and thalassaemia-related bone disease c. Eye problems, including sickle retinopathy and chelation-related eye disease d. Cardiological complications, including sickle cardiomyopathy and iron overload related heart disease e. Chronic respiratory disease, including sickle lung disease and obstructive sleep apnoea f. Endocrine and growth problems, including endocrinopathies and osteoporosis g. Neurological complications, including sickle vasculopathy, other complications requiring neurology or neurosurgical input and access to interventional and neuroradiology h. Hepatobiliary disease, including sickle hepatopathy, viral liver disease and iron overload-related liver disease i. Growth delay / delayed puberty j. Enuresis k. Urological complications, including priapism l. Dental problems	Y	
HC-511	**Anaesthesia and Surgery** Guidelines should be in use covering the care of children and young people with sickle cell disorder and thalassaemia during anaesthesia and surgery.	Y	
HC-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, admission units, clinic and ward areas.	Y	

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-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 an HCC / SHT b. Ensuring all patients are reviewed by a senior haematology decision-maker within 14 hours of acute admission c. Patient discussion at local multidisciplinary team meetings (QS HC-604) d. Referral of children for TCD screening if not provided locally e. 'Fail-safe' arrangements for ensuring all children and young people have TCD ultrasound when indicated f. Arrangements for liaison with community paediatricians and with schools or colleges g. Follow up of patients who 'were not brought' h. Transfer of care of patients who move to another area, including communication with all haemoglobinopathy services involved with their care before the move and communication and transfer of clinical information to the HCC, SHT, LHT and community services who will be taking over their care i. If applicable, arrangements for coordination of care across hospital sites where key specialties are not located together j. Governance arrangements for providing consultations, assessments and therapeutic interventions virtually, in the home or in informal locations	Y	

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-603	**Shared Care Agreement with LHTs** A written agreement should be in place with each LHT covering: a. Whether or not annual reviews are delegated to the LHT b. New patient and annual review guidelines (QS HC-502) (if annual reviews are delegated) c. LHT management and referral guidelines (QS HC-503) d. National Haemoglobinopathy Registry data collection (QS HC-701) e. Two-way communication of patient information between HCC / SHT and LHT f. Attendance at HCC business meetings (HC-607) (if applicable) g. Participation in HCC-agreed audits (HC-706)	N	Shared care agreements with LHTs were not in place. All of the 10 LHTS had a low prevalence of patients with a haemoglobin disorder.
HC-604	**Local Multidisciplinary Meetings** MDT 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, when required, representatives of support services (QS HC-301).	Y	
HC-606	**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 children and young people with haemoglobin disorders b. Two-way exchange of information between hospital and community services	n/a	The acute and community team were integrated
HC-607S	**HCC Business Meeting Attendance (SCD)** At least one representative of the team should attend each SCD HCC Business Meeting (QS HC-702).	Y	
HC-607T	**HCC Business Meeting Attendance (Th)** At least one representative of the team should attend each Thalassaemia HCC Business Meeting (QS HC-702).	Y	
HC-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, identify issues of mutual concern and agree action.	Y	

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-701	**National Haemoglobinopathy Registry** Data on all patients should be entered into the National Haemoglobinopathy Registry. Data should include annual updates, serious adverse events, pregnancies, patients lost to follow up and the number of patients who have asked to have their name removed.	Y	
HC-705	**Other Audits** Clinical audits covering the following areas should have been undertaken within the last two years: a. The patient pathway for patients needing regular transfusion, including availability of out-of-hours services and achievement of expected maximum waiting times for phlebotomy, cannulation and setting up the transfusion (QS HC-505) b. Acute admissions to inappropriate settings, including feedback from children, young people and their families and clinical feedback on these admissions	Y	The team had an extensive and active audit programme.
HC-706	**HCC Audits** The service should participate in agreed HCC-specified audits (QS HC-702d).	Y	
HC-707	**Research** The service should actively participate in HCC-agreed research trials.	Y	
HC-797	**Review of Patient Experience and Clinical Outcomes** The service's multidisciplinary team, with patient and carer representatives, should review at least annually: a. Achievement of Quality Dashboard metrics compared with other services b. Achievement of Patient Survey results (QS HC-197) compared with other services c. Results of audits (QS HC-705): i. Timescales and pathway for regular transfusions ii. Patients admitted to inappropriate settings Where necessary, actions to improve access, patient experience and clinical outcomes should be agreed. Implementation of these actions should be monitored.	Y	
HC-798	**Review and Learning** The service should have appropriate multidisciplinary arrangements for review of, and implementing learning from, positive feedback, complaints, serious adverse events, incidents and 'near misses'.	Y	

Ref.	Quality Standards (Children)	Met? Y/ N/	Reviewer Comment
HC-799	**Document Control** All information for children, young people and their families, policies, procedures and guidelines should comply with Trust (or equivalent) document control procedures.	Y	

Return to Index