Care of People with Inherited and Acquired Haemophilia and other Bleeding Disorders

Royal Manchester Children's Hospital

Visit Date: 21st November 2019

Report Date: March 2020

8831

Contents

Introduction

This report presents the findings of the peer review of services for people with Inherited and Acquired Haemophilia and other Bleeding Disorders at the Royal Manchester Children's Hospital which took place on the 21st November 2019.

The purpose of the visit was to review compliance with the Quality Standards for Inherited and Acquired Haemophilia and other Bleeding Disorders (V1 July 2018) which were developed by the UK Haemophilia Centres Doctors' Organisation (UKHCDO) Peer Review Working Group working with the Quality Review Service (QRS).

The peer review visit was organised by QRS on behalf of the UKHCDO.

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 and Annual Governance Statement. 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.

Appendix 1 lists the visiting team and **Appendix 2** gives 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:

- Royal Manchester Children's Hospital
- NHS England Specialised Commissioning North West

Most of the issues identified by quality reviews can be resolved by providers' and commissioners' own governance arrangements. 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 for monitoring their implementation, liaising, as appropriate, with other commissioners.

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

Quality Review Service would like to thank the staff of the Manchester Children's Comprehensive Care Centre for all 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 parents who took time to meet the review team. Thanks, are also due to the visiting team (**Appendix 1**) and their employing organisations for the time and expertise they contributed to this review.

Manchester Comprehensive Care Centre

The Royal Manchester Children's Hospital (RMCH) was part of the multi-hospital complex of Manchester University NHS Foundation Trust and, at the time of the review, was one of the largest paediatric hospitals in Europe with a catchment area of almost one million children under 16 years of age.

The RMCH non-malignant haematology unit formed part of the haematology/oncology department. The unit and staff provided care for patients with bleeding disorders, thrombosis and haemoglobinopathy. The Centre was designated as a Comprehensive Care Centre (CCC) and was one of the largest paediatric haemophilia units. The catchment area stretched down to Leighton (Crewe) on the southern border and into the Lake District in the north. Support was also provided to linked hospitals in Blackburn, Burnley, Lancaster, Preston, Oldham, and Leighton, and to their community teams. In addition to clinics held at RMCH, quarterly clinics were also held at Blackburn/Burnley and Leighton.

The CCC provided treatment, routine care and support for 73 severe, 14 moderate and 42 mild haemophilia patients, (with 69 severe and 6 moderate patients on a prophylaxis regime). There were 207 von Willebrand (vWD) patients and 200 other patients with rare bleeding disorders (including three severe rare diseases with patients on regular therapy). Patients who were 16 years old were kept under the care of the paediatric team until completion of their GCSEs when they were transferred to the adult services. Teenagers were seen in transition clinics, alongside the adult team, once per year.

Access to the unit was via the dedicated haematology/oncology outpatient department. The outpatient department had a large reception area, nine clinic consulting rooms, consultant offices, a dedicated notes library, a well-equipped children's play area and a child friendly waiting room area. Ward 84 consisted of a 29 bed inpatient unit, a 7 bed bone marrow transplant unit, a 4 bed stem cell therapy unit, and a day- case ward with six beds and nine treatment chairs. For families requiring accommodation, the Centre was able to provide the option of The Ronald McDonald House. This facility was offered to families who lived some distance away, if their child required an inpatient stay.

The department had six consultant paediatric haematologists, one of which was an Honorary Professor of Paediatric Haematology. Four of the paediatric haematologists also covered the malignant and Bone Marrow Transplant (BMT) and two covered the non-malignant service. Haematology specialist trainees rotated six-monthly through paediatric haematology and spent three months with the non-malignant service.

The haematology team had been through a challenging period following a number of retirements within the last two years. However, the team had continued to conduct a weekly multidisciplinary haemophilia clinic on Monday mornings from 9am until 1pm. This clinic was held within the designated Ward 84 outpatient department, under the supervision of the specialist haemophilia nurse. Monthly Monday clinic dates were set aside for patients with severe haemophilia and the team had been able to offer some of these patients new therapies in the form of clinical trials, enhanced half-life products, and more recently sub-cutaneous treatment. These treatments with their potential to improve the quality of life for these patients had been introduced with close management and supervision and had been very well received by patients. Additional clinics were set aside on Monday afternoon for new patient referrals, Thursday morning for pre-op clinic, and Friday morning for Idiopathic Thromboctytopenic Purpura (ITP) patients.

The team held weekly multidisciplinary team meetings (MDT) and that followed a set agenda, with inpatients, patients of note from clinics, Paediatric Emergency Department (PED) attendances, planned surgeries, and research issues discussed and actioned. In addition to these meetings, there were monthly joint MDT meetings held at the adult centre with the adult service, with discussions on outcomes and feedback on the teenage transfer clinics. A monthly joint radiology MDT meeting was also incorporated into Thursday afternoon meetings.

The research team had conducted a number of clinical trials in recent years with highly successful outcomes. In 2017 the team recruited the first global patient to the BAX855 Haemophilia A Previously Untreated Patients (PUP) study. This resulted in Baxalta obtaining approval for the drug. This was recognised as a major achievement in which RMCH had played a significant role. As a result of the success of the BAX clinical trials, RMCH was chosen as part of a Medicines and Healthcare products (MHRA) inspection for three BAX 855 Extended Half Life products haemophilia A clinical trials. The inspection was carried out with no major findings.

Nursery/school visits and home visits for newly diagnosed haemophilia patients were provided by the haemophilia specialist nurse. In addition, training had been completed at District General Hospitals and with some community teams to deliver a specialist service tailored to the patients' needs. This was a vital service for those patients living some distance away, or with significant social needs.

The team provided training for families to enable them to become involved and administer treatment at home. This training was offered to the families of severe haemophilia patients and patients with moderate haemophilia who required regular treatment. Patients receiving the home treatment delivery service were required to use Haemtrack to record their treatments. RMCH was involved with the Haemtrack monitoring CQUIN, which aimed to achieve a compliance rate of 85% over a three-year period. At the time of the visit, compliance was 88%.

Laboratory medicine was split, with paediatric histopathology situated in RMCH and other paediatric laboratory sub-specialties in the Clinical Science Buildings that served the multi-hospital complex. Molecular biology facilities were shared.

At the time of the review, the following numbers of patients were registered at the Centre:

Condition		No. patients	Number of patients who had an annual review in last year	Number of inpatient admissions in last year
Haemophilia A		Total = 114 64 Severe 10 Moderate 40 Mild	104 *64 (60 had 6-monthly reviews)* *10* *30*	22
Haemophilia B		Total = 15 9 Severe 4 Moderate 2 Mild	15 9 *(8 had 6-monthly reviews)* 4 2	1
Von Willebrand		Total = 207 12 Type 3	180 *10 (had 6-monthly reviews)*	2
Other	 FV FVII FX FXI FXIII Glanzmann's Bernard Soulier Storage Pool	Total = 223 7 72 8 57 8 2 4 39	184 *All patients were offered annual reviews. Severe patients were offered 6-monthly reviews*	4

Emergency Care

All patients had access to the Paediatric Emergency Department (PED) and were advised to seek advice there. Guidelines for the care of children with Inherited and Acquired Bleeding Disorders were located in the PED, the Ward 84 doctor's office, Ward 84, and the haematology consultant offices.

Ward Care

Ward 84 had 29 beds for haematology/oncology inpatients. The Bone Marrow Transplant Unit (BMTU) had seven inpatient beds and the stem cell unit had four inpatient beds. The inpatient units were fully established and open 24/7.

Day Care

There were six beds and nine chairs for day case admissions for haematology / oncology patients. This facility was open Monday to Friday 8am to 8pm.

Outpatient Care

A dedicated haematology /oncology outpatient clinic with full facilities was available, it included nine consulting rooms, a child friendly waiting area and play area, toilet facilities and baby changing facilities. This was open Monday to Friday 8am to 6pm.

Community Based Care

The team provided education and training for parents so that they could become involved with and help to provide care for their child. This service was offered to families of severe haemophilia and moderate haemophilia patients and those requiring regular treatment. The training enabled them to administer the child's treatment at home. Nursery/school visits were provided by the haemophilia specialist nurse as were home visits for newly diagnosed haemophilia patients.

In addition, training had been provided at District General Hospitals and also with some community teams, where vital support had been provided in special circumstances.

Return to Index

Review Findings

Achievements

This Comprehensive Care Centre team was enthusiastic and cohesive, working very hard and offering a good service. The relatively new senior team were making strides towards important service improvements, including gaining approval for a business case to appoint much needed additional staff in the non-malignant haematology team (see Concern 1 below). They were achieving a great deal despite their current staffing limitations. The contribution of all team members was noted, with that of the senior nurse specialist and the social worker being outstanding. Play support was also strong.

There was an active research portfolio, which the team was keen to expand. The haemophilia team were supported by, and had strong working relationships with, the hospital research team, especially nurses.

The facilities from which the service was offered were of a very high standard: bright and child-friendly, with a large day care area and a 'crash room' [1]on the in-patient ward for the use of teenagers.

Good Practice

1. Multi-disciplinary team working was strong, supported by clear terms of reference, and there were monthly joint MDT meetings with the adult CCC team at the Manchester Royal Infirmary (MRI). Monthly meetings were also held jointly with radiology.
2. Several guidance documents were noteworthy including: a 'home care charter'; a guideline on factor ordering, storage and stock control; a parent and carer competency document for children on home treatment; a letter to families from the social worker outlining what she could offer; and an excellent detailed individual healthcare plan for schools which included multiple carer contacts, indicating who could give factor replacement and so could most usefully be called if the child had an accident or bleed while at school.
3. The guideline for children undergoing surgery, and the surgical management plans, were very good and the team held a 'virtual' clinic weekly to discuss plans for children having any procedures in the forthcoming week.
4. There had recently been a heightened focus on the transition process, with members of the children's and adult team taking part in an NHS improvement programme, and at the time of the review there was a joint monthly transition clinic held at the adult centre for young people from the age of 14 upwards. It was understood that the monthly clinic, an increase from the previous twice-yearly transfer clinic, was still in pilot form; reviewers considered to be a priority that this should continue.
5. The dental team worked in the severe bleeding disorders monthly clinic and were able to see children regularly without them having to attend for separate appointments.
6. When clinical advice had been given to linked District General Hospitals regarding children who had presented there, an e-mail outlining the question was requested and a response summarised the advice given, for the patient record.
7. There was a dedicated treatment room for haemophilia with a novel '3D television' in the treatment room providing good distraction for children having venepunctures or other interventions. This room was available all day and every day, for use by the nursing staff during scheduled clinics and for out of clinic and emergency attenders.

[1] A room where teenagers were able to relax, with facilities to play games etc.

Immediate Risks

There were no immediate risks identified at the time of the visit.

Concerns

1. **Staffing**[2]

    a. Senior medical staff sessions - Two consultant specialists had a total of 0.8 WTE devoted to this large regional specialist service, and there was no time in the Centre director's job plan for this particular role and no time to undertake management work underpinning the service (for example developing and refining clinical guidelines). There were also considerable time pressures within clinics, with long waiting times for patients and families.

    b. Physiotherapy - Within the 0.2 WTE allocated to the service, a Band 7 physiotherapist could offer only a reactive service, with no time to work with families on early recognition of joint bleeds, other education, or advice about activities. Although she attended the monthly clinics for severely affected children (with the number of children attending being up to fifteen in a single morning) and all patients who attended were seen by a physiotherapist, there was often not time to undertake a full consultation. Children with mild and moderate disorders were seen only if there was a specific reason, so they did not benefit from any musculoskeletal education or advice.

    c. Nursing - Given the large number of patients, residing across a wide geographical area, specialist nursing provision was not adequate, and it had become impossible to offer school visits for many children. The situation was exacerbated by long-term sickness in the wider non-malignant haematology nursing team. The provision, and grading, of the specialist nurses should be reviewed.

    d. Psychology - There was no psychologist working within the team, although this is an expected core team member for patients with these conditions. Patients could be referred to the general hospital psychology service, but waiting times were long. Under these circumstances, only the highest-level patient and family needs were being met, and the team lacked the professional support that an integrated psychologist team member brings.

2. **Guidelines**

    The diagnostic and clinical guidelines needed to be revised. Many were in the form of national published guidance, without any indication as to their local application in the Centre setting; some were locally written but two different versions were seen. Reviewers noted the following examples where revisions were required: in the guidelines on the management of acute joint bleeds, the optimal factor level to aim for was included[3], but not guidance on how to calculate the dose required; in a von Willebrand's disease guideline there was no specific indication of fluid volumes permissible when administering DDAVP[4] and a

[2] Following the peer review, it was confirmed that a business case had been approved for the recruitment of an additional consultant for haemoglobinopathy, an additional 0.5 Band 6 nurse for haemostasis and some secretarial support. Further nurses will also be appointed into the Haemoglobinopathy team which will help with the cross-cover pressures for members of the haemophilia team.

[3] The Centre have since confirmed that dose calculation guidance was included but agreed that this was not clearly signposted.

[4] **DDAVP** - Desmopressin (also known as DDAVP**,** which stands for 1-deamino-8-D-arginine vasopressin) is a synthetic medicine that boosts levels of factor VIII (FVIII) and von Willebrand factor (VWF) to prevent or control bleeding.

guideline on home therapy was judged not to be sufficiently detailed or practical. Most of the locally written guidelines were in draft form, and it was not clear how they were currently accessed as they were not available on the Trust's intranet.

3. **Paediatric Emergency Department (PED)**

   There was no alert on the electronic patient record to indicate that a child had a bleeding disorder, and no indication on the summary care plan or in other guidance to avoid the child being given giving intramuscular injections (IMI) or non-steroidal anti-inflammatory agents (NSAIDs). An audit had demonstrated delays in children receiving treatment on presentation to the PED; this was the focus of some ongoing work. There were two versions of the guideline (one for triage and one for doctors) however it was unclear which staff group each applied to, which could lead to confusion. In addition, a flow-chart displayed in the PED was not included in the PED guideline and did not appear to be available on the intranet. A document entitled 'Management of acute bleeding episodes', written for new trainee medical staff, was found to be the most useful and practical and could form the basis of a re-written guideline for PED staff.

Further Consideration

1. Condition-specific information was sparse, and very little was seen on the rarer conditions even though many children with rarer disorders were managed at the Centre. Although transition practice was good, after recent improvement activities, there was not yet any information for young people preparing for, and going through, the process.
2. Families reported that it was not always easy to access the team by phone. The primary number given was a landline to a phone in the nurses' office; if this could not be answered, calls were relayed to administrative and clerical staff. A direct generic e-mail address to which non-urgent queries could be sent would also be much appreciated by parents and carers.
3. The two specialist haemostasis and thrombosis (HT) consultants took part in a 1:6 out-of-hours on-call rota. If the consultant on call was one of the other four paediatric haematologists, whose interests were in malignant haematology, they contacted one or other of the HT consultants informally for guidance on children with complex bleeding presentations. A formal '2nd on call' arrangement would be more robust.
4. Discussions about potential or obligate female relatives, who should be recalled for testing and counselling before they reach reproductive age, did form part of clinic consultations with families, and with patients in the adult service. However, there was no robust, fail safe system in place for identifying and recalling these young women. A joint policy between the adult and paediatric comprehensive care centres would be useful in order to ensure that identifiable young women carriers were tested or counselled before they became pregnant.
5. The operational policy indicated that if children were not brought for two or more appointments, they would be discharged. In practice, social services were involved if children were not being brought and the policy should reflect this necessary referral. Some of the content of the operational policy still referred to the haemoglobin disorder policy, from which it appeared to have been derived. A review to delete these sections would be useful.
6. Recent audit, for example of the PED pathway, had yielded important outcomes that were being addressed and there were now plans in place for a wider programme of audit of different aspects of the service. This work should be prioritised to identify any other aspects of care that required the same sort of attention and improvement work.
7. Nurse-led telephone clinics for children with milder bleeding disorders had been discussed, but not yet started. These clinics are highly valued by families, saving long journeys - often for brief consultations - for those living distantly, and helping to reduce clinic waits for other attenders.

8. Haemtrack usage was good, but it appeared that the team were not always taking the opportunity to follow up when a family entered data reporting a bleed and a need for home treatment.
9. The parents who met reviewers suggested that a support group, especially for the parents of newly diagnosed children, would be much appreciated by families and carers.
10. Although FEIBA was not held or used in the Centre at the time of the review, the next iteration of the clinical guideline could usefully include an alert that patients on Emicizumab should not be given this product if presenting with a bleed, because of the risk of thrombotic complications resulting from the two agents in combination.
11. The current electronic patient records were not accessible by consultants on call from home out of hours, and any planned upgrade should ensure this facility is available.
12. Document control was not complete, with many guidelines being in draft, or without details of authorship, approval date and planned review date. The name of the previous Centre director still appeared in some guidance.
13. Signage to the Centre was poor, with essentially no indication that this large specialist service was provided alongside malignant haematology services.

Return to Index

General Comment

There was no formal network in place, as there were no haemophilia centres linked with the CCC at RMCH. However, a functional informal network existed, with the CCC team offering advice to six linked District General Hospitals, at Blackburn, Burnley, Lancaster, Preston, Oldham and Leighton, and despite sessional pressures the consultants were managing to undertake a total of eight outreach clinics at these sites per year. There were plans for the Centre leads to meet up in the near future with potential link paediatricians in these hospitals, to progress joint working in the form of the sharing of guidelines, clinical audits, and shared educational opportunities. The job plans of the two consultants at the CCC will need to be reviewed if they are to undertake this development work. It is hoped that this will be possible, and further PA's committed to the service, when an appointment is made to the additional non-malignant haematology consultant post; this appointment was approved but not finally signed off at the time of the visit.

Discussions were starting with the team at the Alder Hey Children's Hospital NHS Foundation Trust where the CCC for Merseyside and parts of Cheshire, Lancashire, Shropshire, North Wales and the Isle of Man was centred. Commissioners were reported not yet to have taken an active role in these discussions, although they were invited to meetings. Active commissioner engagement is likely to be key to any progress resulting from these meetings.

Return to Index

APPENDIX 1 Membership of Visiting Team

Visiting Team		
Leah Denver	Data Manager Haemophilia Unit	Birmingham Women's and Children's NHS Foundation Trust
Nicola Hubert	Paediatric Physiotherapist	Great Ormond Street Hospital for Children NHS Foundation Trust
Dr Anne Kelly	Consultant Haematologist	Cambridge University Hospitals NHS Foundation Trust
Janice Llewellyn	Haemophilia Nurse (Paeds)	The Shrewsbury and Telford Hospital NHS Trust
Clive Smith	Patient representative	Chair, Haemophilia Society

QRS Team		
Rachael Blackburn	Assistant Director	Quality Review Service
Dr Anne Yardumian	Consultant Haematologist	Programme Clinical Lead

Return to Index

APPENDIX 2 Compliance with the Quality Standards

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

Table 1 - Percentage of Quality Standards met

Details of compliance with individual Quality Standards can be found below.

Service	Number of applicable QS	Number of QS met	% met
Centre	37	23	62
Network	8	7	88
Commissioning	3	3	100
Total	**48**	**33**	**69**

Return to Index

Comprehensive Care Centre

Ref	Standard	Met?	Comments
HP-101	**Service Information** Written information should be offered to patients and, where appropriate, their carers covering at least: a. Brief description of the service 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 HP-199)	Y	

Ref	Standard	Met?	Comments
HP-102	**Condition-Specific Information** Written information should be offered to patients and, where appropriate, their carers covering at least: a. A description of their condition and how it might affect them b. How their condition is diagnosed c. Genetics of inherited bleeding disorders d. Testing for carrier status and the implications of being a carrier e. Problems, symptoms and signs for which emergency advice should be sought f. Out of hours services g. 'On demand' clotting factor treatment h. Prophylaxis i. Self infusion (or infusion by parent or carer) j. Home therapy and use of Haemtrack k. How to manage bleeding at home l. Ports, fistulae and in-dwelling access devices (if applicable) m. Possible complications, including inhibitors and long term joint damage n. Approach to elective and emergency surgery o. Women's health issues p. Health promotion, including smoking cessation, health eating, weight management, exercise, alcohol use, sexual and reproductive health, and mental and emotional health and well-being q. Dental care r. Travel advice s. Vaccination advice t. National Haemophilia Database, its purpose and benefits u. Sources of further advice and information Information should be available covering: 1. Haemophilia A 2. Haemophilia B 3. Von Willebrand Disease 4. Acquired haemophilia 5. Inherited platelet disorders 6. Other less common and rare bleeding disorders	N	Condition-specific information was sparse, and very little information was seen on the rarer conditions even though many children with rarer disorders were managed at the Centre.

Ref	Standard	Met?	Comments
HP-103	**Plan of Care** Each patient and, where appropriate, their carer should discuss and agree their Plan of Care, and should be offered a written record covering at least: a. Agreed goals, including life-style goals b. Self-management c. Planned assessments, therapeutic and/or rehabilitation interventions d. Early warning signs of problems, including acute exacerbations, and what to do if these occur e. Agreed arrangements with school or other education provider and preparation for adult life (children and young people only) f. Planned review date and how to access a review more quickly, if necessary g. Who to contact with queries or for advice The Plan of Care should be communicated to the patient's GP and to relevant other services involved in their care.	Y	
HP-104	**Review of Plan of Care** A formal review of the patient's Plan of Care should take place at least six monthly for patients with severe or moderate haemophilia and at least annually for other patients. This review should involve the patient, where appropriate their carer, and appropriate members of the multi-disciplinary team. Haemtrack results should be reviewed (if applicable) and the outcome of the review should be communicated in writing to the patient and their GP.	Y	
HP-105	**Contact for Queries and Advice** Each patient and, where appropriate, their carer should have a contact point within the service for queries and advice. A clear system for triage of urgent clinical problems should be in place. If advice and support is not immediately available for non-urgent enquiries, then the timescales for a response should be clear. Response times should be no longer than the end of the next working day. All contacts for advice and a sample of actual response time should be documented.	Y	
HP-106	**Haemtrack (Patients on Home Therapy)** All patients on home treatment should be encouraged to use electronic recording of their treatment through Haemtrack.	Y	

Ref	Standard	Met?	Comments
HP-194	**Environment** The environment and facilities in out-patient clinics, wards and day units should be appropriate for the number of patients with inherited and acquired bleeding disorders and accessible by people with severe mobility problems. 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	
HP-195	**Transition to Adult Services and Preparation for Adult Life** 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 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 afterwards f. Advice for young people going away from home to study, including: i. registering with a GP ii. how to access emergency and routine care iii. how to access support from their Comprehensive Care Centre iv. communication with their new GP	N	Although transition practice had been developed and was good, information for patients was not in place.
HP-198	**Carers' Needs** Carers should be offered information on: a. How to access an assessment of their own needs b. What to do in an emergency c. Services available to provide support	N	Reviewers did not see any evidence that parents' / carers' needs were assessed.
HP-199	**Involving Patients and Carers** The service should have: a. Mechanisms for receiving regular feedback from patients and carers about treatment and care they receive b. Mechanisms for involving patients and carers in decisions about the organisation of the service c. Examples of changes made as a result of feedback and involvement of patients and carers	N	Reviewers did see some patient feedback, but this was limited and there was no evidence for 'b' or 'c'.

Ref	Standard	Met?	Comments
HP-201	**Lead Consultant and Lead Nurse** A nominated lead consultant and lead nurse should have responsibility for staffing, training, guidelines and protocols, service organisation, governance and for liaison with other services. The lead consultant and lead nurse should be registered healthcare professionals with appropriate specialist competences and should undertake regular clinical work within the service and specific time allocated for their leadership role.	Y	
HP-202	**Staffing Levels and Skill Mix** Sufficient staff with appropriate competences should be available for out-patient, day unit and in-patient care and for support to urgent care services. Staffing levels should be appropriate for the number of patients cared for by the service and its role in the network. All staff should undertake regular Continuing Professional Development of relevance to their work in the inherited and acquired bleeding disorders services. Staff working with children and young people should have competences in caring for children as well as in the care of people with bleeding disorders. Cover for absences should be available. In HCCCs these staff should have sessional time allocated to their work with the IABD service. In HCs the arrangements for accessing staff who do not have sessional time allocated to the IABD service should be clearly defined. Staffing should include: a. Medical staff: i. Consultant specialising in the care of people with inherited and acquired bleeding disorders available during normal working hours ii. On-call consultant haematologist (24/7) iii. Arrangements for advice from a consultant specialising the care of people with inherited and acquired bleeding disorders (if not on call) b. Specialist nursing staff: i. Bleeding disorders specialist nurses (5/7) ii. Ward, out-patient and day unit staff with competences in the care of people with inherited and acquired bleeding disorders. c. Clinical specialist physiotherapist d. Biomedical Scientist and Clinical Scientist (further detail on the requirements are included in HP-303) e. Clinical or counselling specialist psychologist or appropriately trained psychotherapist f. Specialist senior social worker g. Data manager	N	See Concerns section of main report.

Ref	Standard	Met?	Comments
HP-203	**Service Competences and Training Plan** The competences expected for each role in the service should be identified. A training and development plan for achieving and maintaining competences should be in place.	N	Evidence was seen for members of the nursing team but not for other members of the wider MDT.
HP-204	**Competences – All Health and Social Care Professionals** All health and social care professionals working in the service should have competences appropriate to their role in: a. Safeguarding children and/or vulnerable adults b. Recognising and meeting the needs of vulnerable children and/or adults c. Dealing with challenging behaviour, violence and aggression d. Mental Capacity Act and Deprivation of Liberty Safeguards e. Resuscitation	Y	
HP-299	**Administrative, Clerical and Data Collection Support** Administrative, clerical and data collection support should be available.	Y	
HP-301	**Support Services** Timely access to the following support services should be available: a. Play support (children's services only) including: i. Play and distraction during any painful or invasive procedures ii. Play support to enable the child's development and well-being b. Pharmacy c. Dietetics d. Occupational Therapy e. Orthotics	N	Evidence was not seen for 'b'.
HP-302	**Emergency Department – Staff Competences** Medical and nursing staff working in the Emergency Department should have competences in urgent care of people with inherited and acquired bleeding disorders including awareness of: a. Guidelines on care of patients with inherited and acquired bleeding disorders in the Emergency Department (QS HP-504) b. Who to contact for advice	N	Evidence was not seen for this Quality Standard and reviewers heard that ED training was not being delivered by the haemophilia team at the time of the review.

Ref	Standard	Met?	Comments
HP-303	**Laboratory Service** a. A UKAS / CPA accredited laboratory service with satisfactory External Quality Assurance performance should be available 24/7 b. A laboratory representative (senior biomedical scientist or clinical scientist) should attend inherited and acquired bleeding disorder service multi-disciplinary team meetings (QS HP-602) regularly c. The following tests should be available: i. All coagulation factor assays (24/7) ii. Inhibitor screening iii. FVIII inhibitor quantification iv. VWF antigen v. VWF activity vi. Platelet function testing d. Molecular Genetic Laboratory service for: i. detection of causative mutations in patients with inherited bleeding disorders ii. carrier detection	Y	
HP-304	**Specialist Services** Timely access to the following specialist staff and services should be available as part of a HCCC service. HCs should be able to access these services through network arrangements: a. Obstetrics including reproductive counselling, information about pre-implantation genetic diagnosis and antenatal diagnosis b. Foetal medicine c. Vascular access (consultant surgeon or interventional radiologist with experience of venous access devices) d. Orthopaedic surgery e. Care of older people services f. Dental services g. HIV services h. Hepatology i. Medical genetics (Genetic Counselling Services) j. Pain management services k. Rheumatology Specialist services should have an appropriate level of specialist expertise in the care of people with inherited and acquired bleeding disorders.	Y	

Ref	Standard	Met?	Comments
HP-402	**Facilities and Equipment** Facilities and equipment appropriate for the service provided should be available including: a. Fridges b. Storage c. Clinical rooms for staff of all disciplines to see patients and carers d. Room for multi-disciplinary discussion e. Room for educational work with patients and carers f. Office space for staff g. Access to Haemtrack and the Haemophilia Centre Information System (HCIS) in all clinical areas All equipment should be appropriately checked and maintained.	Y	
HP-499	**IT System** IT systems should be in use for: a. Storage, retrieval and transmission of patient information, including access to the latest vCJD status and family tree b. Patient administration, clinical records and outcome information c. Data to support service improvement, audit and revalidation d. Alerting the specialist team when patients attend the Emergency Department	N	Evidence was not seen for 'a', 'c', or 'd'.
HP-501	**Diagnosis Guidelines for Patients with Suspected Inherited and Acquired Bleeding Disorders** Guidelines on diagnosis should be in use covering at least a. Haemophilia A b. Haemophilia B c. Von Willebrand Disease d. Acquired haemophilia e. Inherited platelet disorders f. Other less common and rare bleeding disorders	N	National guidelines were in place, but they had not all been localised (see Concerns section of main report). 'd' is not applicable.

Ref	Standard	Met?	Comments
HP-502	**Guidelines: Concentrate Use and Monitoring** Guidelines should be in use covering: a. Concentrate therapy: i. Initiation and monitoring of prophylaxis ii. Home therapy b. Use of extended half life products, including inhibitor testing and PK assessment c. Management of concentrate supplies including: i. Ordering ii. Storage iii. Stock control to ensure all stock is up to date and waste is minimised iv. Prescription and delivery for patients on home treatment v. Arrangements for emergency 'out of hours' supply vi. Recording issue to patients vii. Recording use by patients, including on Haemtrack viii. Submission of data via NHD for national tenders coordinated by CMU	Y	
HP-503	**Clinical Guidelines** The following clinical guidelines should be in use: a. Management of acute bleeding episodes, including patients with inhibitors b. Inhibitor screening c. Immune tolerance therapy d. Dental care e. Care of patients with hepatitis C f. Care of patients with HIV g. Antenatal care, delivery and care of the neonate h. Management of synovitis and target joints i. Long term surveillance of musculoskeletal health j. “For public health purposes”: care of patients at risk of vCJD who are undergoing surgery	N	National guidelines were in place, but they had not all been localised (see Concerns section of main report).
HP-504	**Emergency Department Guidelines** Guidelines on management of patients with inherited and acquired bleeding disorders in the Emergency Department should be in use.	Y	

Ref	Standard	Met?	Comments
HP-505	**Guidelines on Care of Patients requiring Surgery** Guidelines on the care of patients with inherited and acquired bleeding disorders who require surgery should be in use covering at least: a. Involvement of surgical and inherited and acquired bleeding disorders service in agreement of a written plan of care prior to, during and post-surgery b. Communication of the agreed plan of care to all staff involved in the patient's care prior to, during and post-surgery c. Documentation of care provided d. Arrangements for escalation in the event of unexpected problems	Y	See Good Practice section of main report.
HP-595	**Guidelines on Transition and Preparing for Adult Life** Guidelines on transition of young people from paediatric to adult services should be in use covering at least: a. Taking responsibility for their own care b. Involvement of the young person and, where appropriate, their carer in planning the transfer of care c. Joint meeting between paediatric and adult services in order to plan the transfer d. Allocation of a named coordinator for the transfer of care e. A preparation period prior to transfer f. Arrangements for monitoring during the time immediately after transfer g. Advice for young people going away from home to study, including: i. registering with a GP ii. how to access emergency and routine care iii. how to access support from their Comprehensive Care Centre iv. communication with the young person's new GP	Y	
HP-599	**Care of Vulnerable People** Guidelines for the care of vulnerable children, young people and adults should be in use including: a. Restraint and sedation b. Missing patients c. Mental Capacity Act and the Deprivation of Liberty Safeguards d. Safeguarding e. Information sharing f. Palliative care g. End of life care	Y	

Ref	Standard	Met?	Comments
HP-601	**Service Organisation** The service should have an operational procedure covering at least: a. Ensuring all children who are in-patients have a named consultant paediatrician and a named haematologist with expertise in caring for patients with inherited and acquired bleeding disorders responsible for their care b. Ensuring all adults are under the care of a consultant haematologist with an interest in inherited and acquired bleeding disorders, either directly or through a shared care arrangement with a general haematologist c. Responsibility for giving information and education at each stage of the patient journey d. Arrangements for involving Haemophilia Centre staff in multi-disciplinary discussions relating to their patients (QS HP-602) e. Arrangements for follow up of patients who 'do not attend' f. Arrangements for transfer of patient information when patients move areas temporarily or permanently g. Ensuring patients' plans of care are reviewed at least six monthly for patients with severe haemophilia and at least annually for other patients (QS HP-104) h. Ensuring school visits for children with severe haemophilia at least at each change of school (children's services only) i. Ensuring patients are visited at home at least annually if they are unable to attend clinics, including those in nursing homes j. Lone working	Y	
HP-602	**Multi-Disciplinary Team Meetings** Multi-disciplinary team meetings to discuss patients' plans of care should take place regularly involving: a. All core members of the specialist team (HP-202) b. Senior biomedical scientist or clinical scientist with responsibility for the Coagulation Laboratory c. HC staff who are regularly involved in the patient's care as part of network arrangements	Y	

Ref	Standard	Met?	Comments
HP-603	**Multi-Disciplinary Clinics** The following multi-disciplinary clinic arrangements for patients with inherited and acquired bleeding disorders should be in place: a. Involvement of medical, specialist nursing and physiotherapy staff in clinics b. Availability of social work and psychology staff in clinics c. Combined clinics or other arrangements for multi-disciplinary discussion with: i. orthopaedics ii. rheumatology iii. obstetrics and gynaecology iv. paediatrics v. dental vi. HIV / hepatology	Y	
HP-604	**Liaison with Other Services** Review meetings should be held at least annually with specialist services to consider liaison arrangements and address any problems identified.	N	
HP-701	**Data Collection** The following data should be collected: a. UK National Haemophilia Database data on all patients b. Data on concentrate use and bleeds, either through Haemtrack or an equivalent mechanism c. Data required to complete the UKHCDO National Haemophilia Dashboard or other national mechanisms	Y	
HP-702	**Audit** The services should have a rolling programme of audit covering at least: a. Clinical guidelines (QS HP-503) b. Emergency and out of hours care (QS HP-504) c. Initiation of prophylaxis in children d. Inhibitor surveillance and Immune Tolerance Induction (ITI) e. Clinical reviews including joint scores (QS HP-103 & 104) f. Concentrate use and wastage	N	Although some audit activity was evident it did not cover all the requirements of this Quality Standard.
HP-706	**Research** The service should actively participate in research relating to the care of patients with bleeding disorders.	Y	

Ref	Standard	Met?	Comments
HP-798	**Multi-disciplinary Review and Learning** The service should have multi-disciplinary arrangements for review of and implementing learning from: a. Positive feedback, complaints, outcomes, incidents and 'near misses' b. Morbidity and mortality c. Haemophilia Dashboard d. Review of UKHCDO Annual Report benchmarking information on concentrate use e. Ongoing reviews of service quality, safety and efficiency f. Published scientific research and guidance	Y	
HP-799	**Document Control** All policies, procedures and guidelines should comply with Trust (or equivalent) document control procedures.	N	Evidence of authorship and review dates was not seen for all documentation.

Return to Index

Network

Ref	Standard	Met?	Comments
HY-199	**Involving Patients and Carers** The network should have mechanisms for involving patients and their carers from all services in the work of the network.	Y	
HY-203	**Inherited and Acquired Bleeding Disorders Network Leads** The network should have a nominated: a. Lead consultant and deputy b. Lead specialist nurse c. Lead physiotherapist d. Lead clinical or counselling psychologist e. Lead manager	Y	
HY-204	**Education and Training** The network should have agreed a programme of education and training to help services achieve compliance with QSs HP-203.	Y	
HY-503	**Guidelines** Network guidelines should have been agreed covering: a. Diagnosis for patients with suspected inherited and acquired bleeding disorders (QS HP-501) b. Concentrate use and monitoring (QS HP-502) c. Clinical guidelines (QS HP-503) d. Management of patients with inherited and acquired bleeding disorders in the Emergency Department (QS HP-504) e. Care of patients requiring surgery (QS HP-505) f. Transition and preparing for adult life (QS HP-595)	Y	
HY-701	**Ongoing Monitoring** The network should monitor on a regular basis: a. Submission of data on all patients to the UK National Haemophilia Database (QS HP-701) b. Network-wide data on concentrate use and bleeds	Y	
HY-702	**Audit** The network should have an agreed programme of audit and review covering network-wide achievement of QS HP-702.	N	See HP-702.
HY-703	**Research** The network should have agreed: a. A policy on access to research relating to the care of patients with inherited and acquired bleeding disorders b. A list of research trials available to all patients within the network.	Y	

Ref	Standard	Met?	Comments
HY-798	**Network Review and Learning** Representatives of Comprehensive Care Centres and referring Haemophilia Centres should meet at least once a year to: a. Identify any changes needed to network-wide policies, procedures and guidelines b. Review results of audits undertaken and agree action plans c. Review and agree learning from positive feedback, complaints, critical incidents and 'near misses', including those involving liaison between teams d. Share good practice and potential service improvements	Y	

Return to Index

Commissioning

Ref	Standard	Met?	Comments
HZ-601	**Commissioning of Services** Commissioners should have agreed the configuration of clinical networks including: a. Designated Comprehensive Care Centres and Haemophilia Centres and the relationships between them b. Whether the service cares for children, adults or both c. Referral pattern to each service, taking into account the type of patients who will be treated by each team	Y	
HZ-701	**Clinical Quality Review Meetings** Commissioners should regularly review the quality of care provided by: a. Each service, including achievement of QS HP-701 b. Each network, including achievement of QS HY-701 and QS HY-798 c. Service and network achievement of relevant QSs	Y	
HZ-798	**Network Review and Learning** Commissioners should attend a Network Review and Learning meeting (HY-798) at least once a year for each network in their area.	Y	

Return to Index