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

## University Hospital Southampton NHS Foundation Trust

Visit Date: 9th September 2019

Report Date: December 2019

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## Introduction

This report presents the findings of the peer review of services for people with Inherited and Acquired Haemophilia and other Bleeding Disorders at University Hospital Southampton NHS Foundation Trust which took place on the 9th September 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:

- University Hospital Southampton NHS Foundation Trust
- Specialised Commissioning NHS England South

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 Southampton Comprehensive Care Centre for their work in preparing for the review and for their kindness and helpfulness during the course of the visit. We are grateful too to the patients, parents and carers 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.

## Southampton Comprehensive Care Centre

The Southampton Haemophilia Comprehensive Care Centre (SHCCC) provided care to adult and paediatric patients in South Hampshire, the surrounding District General Hospitals and Guernsey. University Hospital Southampton (UHS) provided services to 1.9 million people within its catchment area and provided specialist services to a catchment of 3.7 million people in southern England and the Channel Islands. Specialist services included Southampton Children's Hospital, obstetrics, orthopaedics, hepatology, genetics, infectious diseases, neurosurgery, cardiac surgery and trauma. At the time of the visit, the haemophilia team comprised three consultants (with one of these specialising in research)[1] who all specialised in haemostasis, two adult and paediatric trained haemophilia nurse specialists, adult and paediatric physiotherapists, a data manager and coagulation laboratory staff. As this was a specialist Trust, the team were often required to manage bleeding disorder patients having complex surgery from a wide region. This required frequent collaboration with haemophilia specialists from the surrounding hospitals.

SHCCC was based in the children's outpatient department and comprised a nursing office and two treatment rooms. Haemophilia patients had open access to the Centre during working hours, and there was a pathway for patients to access services out of hours via the haematology on-call registrar or consultant.

Bleeding disorder patients in South Hampshire and Dorset had opportunities to participate in research, as the Centre ran a number of haemophilia trials, including gene therapy trials with administration of gene therapy at the National Institute for Health Research (NIHR) Southampton clinical research facility. The research team included a nurse and research fellow dedicated to non-malignant haematology trials.

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

| Condition | | No. patients And breakdown of severity | No. patients who had an annual review in last year | No. in-patient admissions in last year |
|---|---|---|---|---|
| Haemophilia A | Adults | 87 | 50 | 13 |
| | Children | 43 | 26 | 4 |
| Haemophilia B | Adults | 18 | 11 | 4 |
| | Children | 6 | 2 | 0 |
| Von Willebrand | Adults | 110 | 49 | 5 |
| | Children | 38 | | |
| Other | Adults | 210 | 114 | 0 |
| | Children | 58 | | |

[1] The Centre team subsequently confirmed that two paediatric haematologists had been appointed since the review who will look after haemophilia patients.

| Diagnosis | SEVERE | MODERATE | MILD | TOTAL |
|---|---|---|---|---|
| Haemophilia A - Adults | 22 | 6 | 59 | **87** |
| Haemophilia A - Paeds | 24 | 6 | 13 | **43** |
| Haemophilia B - Adults | 3 | 5 | 10 | **18** |
| Haemophilia B - Paeds | 3 | 1 | 2 | **6** |
| Von Willebrand - Adults | 3 | 12 | 95 | **110** |
| Von Willebrand - Paeds | 2 | 4 | 32 | **38** |
| Other Diagnosis - Adults | 4 | 13 | 193 | **210** |
| Other Diagnosis - Paeds | 0 | 0 | 58 | **58** |
| **Total** | **61** | **47** | **462** | **570** |

**Note**: The figures in the table above were supplied by the Centre following the review.

## Emergency Care

All bleeding disorder patients were provided with the haemophilia specialist nurses' telephone numbers and the Trust switchboard telephone number. Business cards, with the nurses contact numbers were provided, along with bleeding disorder cards and leaflets with advice on what to do in an emergency. During working hours patients were advised to telephone the nurses for advice regarding minor joint or muscle bleeding. For moderate and major bleeding, head injuries or neurological symptoms, patients were advised to attend the Emergency Department (ED) and contact the nurses once en route to or at the ED, to ensure that emergency treatment could be provided in the most appropriate setting. Out of hours, patients were advised to call the Trust switchboard and state that they had a bleeding disorder. The switchboard would then put the patient through to the haematology registrar to provide advice along with haemophilia consultant support. Children with haemophilia had open access to the Paediatric Assessment Unit (PAU) during evenings and at the weekend, and the on-call haematology registrar could assess them there.

Haemophilia patients had emergency care plans documented on the electronic document system which could be accessed by any doctor or nurse within the hospital as well as by the on-call haematology registrars and consultants from home via the 'remote access' system. Pathways for emergency care of patients with haemophilia were available in the ED and haemophilia centre, and all haematology registrars had an electronic copy.

## Ward Care

Ward D2 was a twenty-bed ward and was the main ward that non-malignant haematology patients were admitted onto. Haemophilia patients requiring admission who primarily needed to be under another speciality (such as orthopaedics), were admitted here.

## Day Care

Ward C7 was a dedicated haematology day unit that cared for patients with malignant and non-malignant haematological disorders. It was open between 8am and 8pm on weekdays between 8am and 2.30pm on Saturdays. Treatment such as Rituximab[2]- for acquired haemophilia, was administered there.

## Outpatient Care

There were two haemophilia treatments rooms based in the children's outpatient's department that were easy to access from the main hospital entrance. There was a dedicated car parking space for haemophilia patients at the front of the hospital. The larger room had toys for the paediatric patients. The treatment rooms were also used as outpatient clinic rooms for bleeding disorder patients. There was one additional clinic room for bleeding disorder patients based in pathology, which was adjacent to children's outpatients. The haematology outpatient supervisor and clerical officer worked closely with the haemophilia team enabling urgent clinic appointments to be arranged easily and changes in patient appointments communicated directly.

## Community Based Care

The haemophilia nurse specialists provided community care when needed, such as home and school visits. Health Care at Home, based within the hospital, were able to take blood tests from patients at home, and administer some treatments. This had proved to be particularly helpful with frail and palliative care patients.

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[2] **Rituximab –** sold under the brand name Rituxan among others, is a medication used in the management of adult patients with haemophilia A and inhibitors resulting in a marked reduction in inhibitor level and clinical improvement in bleeding.

# Review Findings

The completeness of this review was hampered by a number of issues which are summarised in the table on page 3 of this report.

**Achievements**

This long-serving team was offering good services, and the contributions made by the clinical nurse specialists, physiotherapists, laboratory team and research support team were notable.

Patient feedback about the care they received from the team was very positive, and they were happy with the services they received. They did note some staffing shortages (see Concern 2).

The research portfolio was impressive, and there had been a considerable increase in the number of studies being recruited to over the preceding couple of years. Studies included gene therapy dosing, and phase 1, 2 and 3 clinical trials.

**Good Practice**

1. 'Transition weeks' were held within the Trust, during which young people with long-term conditions moving from paediatric to adult services could visit and familiarise themselves with their new teams and services. Parents reported that the transition process for their teenagers had been well managed.
2. Joint working with a named obstetrician was exemplary. Women with bleeding disorders or who were at risk of having an affected infant, and who were planning pregnancy or were already pregnant, were seen at shared fortnightly clinics and there was careful communication with all other members of the extended team as needed. The named obstetrician was also the lead for maternal medicine and chaired the regional maternal medicine committee. Obstetric colleagues often contacted him by email for clinical advice. A specialist colleague at Southampton worked with him and provided cover.
3. Multi-disciplinary Team (MDT) meetings were held weekly and were minuted; the team were told that the MDT discussion outcomes were included in the electronic patient record.
4. Staff could access electronic records to see results and correspondence remotely when on-call out of hours.
5. There had been considerable turnover in laboratory personnel in the preceding year, and senior staff had invested a good deal of time in training incoming junior biomedical scientists to perform the range of specialist investigations required to support the service at all times.
6. The 'My Health Record' software had recently been made available for patients to access their own results and hospital notes and letters.
7. The 'Progeny' clinical family history data management system was available for accurate recording of family tree and genetic information.
8. Although there were no formal network meetings in place, the clinical nurse specialists met regularly with the other nurse specialists across Wessex, and this was a useful support and information sharing group.

**Immediate Risks**

No immediate risks were identified at the time of the visit.

**Concerns**

1. **Data**

   The review team were shown different numbers for patients treated with the various conditions, and for patients who were severely affected, during the course of the day. It was not clear whether the numbers given were for all patients registered at the Centre, or patients under active follow up. There was a degree of ambiguity regarding dual registration of patients and therefore which patients were the sole responsibility of the Southampton team, which made some activity figures, for example the numbers of severely affected patients who had an annual review undertaken within the last year, difficult to evaluate. There had been a problem with serial non-attendance of patients to clinic visits, and it was possible that some patients had been lost to follow up; it was not clear if this could include children – although it was noted that severely affected children were highly unlikely to have been lost to follow up. The lack of a robust denominator of patient numbers made it difficult to assess coverage of some quality indicators, such as the numbers of severely affected patients on prophylactic factor replacement therapy, the number having had six-monthly assessments, and the number of patients having joint scores undertaken by the physiotherapists.

2. **Staffing**

   All staff in post were working very hard to try to ensure that their patients had appropriate care; however, staffing shortfalls were hindering the team's ability to deliver the expected, rounded multi-professional care. Patients especially noted the lack of psychology support and commented on how stretched the nurse specialists were.

   a. Paediatric senior medical staff: At the time of the visit, there was no paediatrician or paediatric haematologist working regularly with the team to deliver specialist care to children with bleeding disorders, and it was not possible for the review team to meet any paediatric medical staff to understand the workings of this service in context of the other paediatric clinical services. Reviewers were informed that a locum paediatric haematologist was due to start very soon, possibly within a month of the visit. However, there was uncertainty about how much time dedicated to this service the consultant was to have in their job plan (please see footnote on page 7).

   b. Physiotherapy: There were adult and paediatric physiotherapists working within the service, but respectively with only one session per month and 3.5 hours per week. Within this time, they were not even able to attend all clinic appointments and were not able to see patients presenting with acute joint bleeds – sometimes for several days. The UKHCDO physiotherapy standards require physiotherapists to have sufficient time and flexibility in their work to see these patients promptly. Much of the physiotherapists dedicated time was spent undertaking formal joint scoring rather than working to promote long term joint health. There were no clinical guidelines relating to managing synovitis, or target joints; it was noted that having guidelines in place would allow for audit of practice, to evidence gaps in service and to the need for additional dedicated time.

   c. Psychology: There was no psychologist working with either the paediatric or the adult team. A psychologist is expected to be part of the core team in caring for these conditions. Where there were high-level concerns, referrals were made through the patient's primary care team. However, the team were lacking in the support and guidance that a psychology member can bring to their practice, and it was inevitable that some lower level patient or family needs were not being met.

   d. Social work: There was no named social worker available to support the patients and families, and it was unclear whether the haemophilia team had easy access to Trust-wide social care support. Social services support (e.g. for completing Personal Independence Payment (PIP) applications) is always greatly appreciated by people affected by long term conditions.

e. Nursing: The lead clinical nurse specialist's work was central to the service and was greatly appreciated by her colleagues and patients. However, there was a concern about the impact on the service if she were off work for any reason or decided to retire. Reviewers heard that the nurses also had increasing demands on their time and that they now had insufficient time to fully support the service in the way they would wish, for example undertaking home visits.

3. **Network**

There was a lack of clarity about a clinical network, specifically in regard to the sites that the Southampton CCC was linked with and supported and about how many adult and paediatric patients were managed at each. The review team heard that home delivery factors were prescribed by the Basingstoke CCC for some patients who had their clinical care at Southampton and who were apparently not also seen by the Basingstoke team. There were pathways in place for paediatric surgery in the region, for which Southampton General Hospital was the regional centre. However, pathways for bleeding disorder management between the hospitals in the area were not clear. One of the Southampton consultant haematologists worked one day per week at Bournemouth; this work was funded through the Southern Haemophilia Network based at Basingstoke. It was understood that the Southampton team planned to offer paediatric clinics at Poole, although the Basingstoke team were already running clinics there every three months.

Urgent discussion with the Southern Haemophilia Network leads and the specialist commissioner (who came to meet the review team and is very engaged with service provision in the region) was required, initially to understand the ways in which the two CCC's (Southampton and Basingstoke) currently operate to provide and support services for patients with bleeding disorders across Wessex, before proceeding to plan clear pathways for the future.

4. **Facilities**

Some of the facilities were of a very high standard, especially in the paediatric areas. However, adults waiting to be seen shared a large impersonal waiting area with people coming for routine blood tests, and those who attended for urgent review waited in a tiny area, not large enough to accommodate a wheelchair. Patient information was not visible in the waiting area and signposting to the Centre from other parts of the hospital was poor. When called through to be seen in a consulting room, patients had to go through the area in which children were waiting for their appointments.

**Further Consideration**

1 Patients reported that their experience of emergency care was poor; ambulance crews sometimes ignored their 'alert card' and even staff in the Emergency Department did not always respond appropriately to the information that they had a significant bleeding disorder, so that assessment and treatment was sometimes delayed.

2 The two consultants with expertise in bleeding disorders were two of five consultants operating a 1:5 adult haematology out of hours rota. If there was a complex presentation during the times they were not on call, their colleagues contacted one or other of them for guidance, on an informal basis. As the haemostasis consultants were also covering the paediatric bleeding disorder service, a formal 'second on call' rota for complex presentations for the two haemostasis consultants would make the process for accessing specialist clinical advice at all times more robust.

3 Some guidelines were not complete, and some were not in place. The diagnostic guidelines lacked sections covering inherited platelet disorders and unusual factor deficiencies e.g. factor XIII. It was noted that these may be important - for example, in assessing a child presenting with bruising that could be due to a bleeding

disorder or to non-accidental injury. There was no operational policy describing and supporting the running of the service.

4 There was no competency framework in place, and there was therefore no evidence that all of the core team members had achieved all the competencies required by their role. In addition, statutory and mandatory training records were not available for all core staff in the Centre.

5 There are some additional investigations which the laboratory may want to add to their repertoire, including measuring Emicizumab levels, human/porcine factor VIII inhibitor screens / levels at weekends (which may be useful for patients presenting with acquired haemophilia), and assays for longer-acting factor replacement agents.

6 The team may want to explore ways of using the Progeny software to put in place a robust system for prompting the recall, for testing and counselling, of potential female carriers of bleeding disorders before they reach reproductive age.

7 Clinic waiting areas could usefully display some of the patient information leaflets that were presented as evidence at the visit.

8 A family from the Isle of Wight requested consideration of an outreach clinic there, for the convenience of a number of patients living there. This might form part of the network development discussions.

9 Document control was incomplete, with some guidelines, and a Trust Policy, lacking authorship or review date.

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# APPENDIX 1 Membership of Visiting Team

| Visiting Team | | |
|---|---|---|
| Emma Franklin | Haemophilia Centre Manager, Adult CNS | University Hospitals Bristol NHS Foundation Trust |
| Sandy Jeffery | Patient representative | |
| Sarah Jones | Specialist Haemophilia Physiotherapist | Cardiff and Vale University Health Board |
| Dr Kate Khair | Consultant Nurse haemophilia | Centre for outcomes and experience research in children's health, illness and disability Great Ormond Street Hospital |
| Dr Susie Shapiro | Consultant Haematologist | Oxford University Hospitals NHS Foundation Trust |
| Dr Oliver Tunstall | Consultant Paediatric Haematologist | University Hospitals Bristol NHS Foundation Trust |

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

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# APPENDIX 2 Compliance with the Quality Standards

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

**Table 1 - Percentage of Quality Standards met**

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

| Service | Number of applicable QS | Number of QS met | % met |
|---|---|---|---|
| Comprehensive Care | 37 | 23* | 62 |
| Network | 8 | 0 | 0 |
| Commissioning | 3 | 2 | 67 |
| **Total** | 48 | 25 | 52 |

* Three Quality Standards (HP 103, HP104 and HP-499) could not be assessed as the Centre did not obtain Caldicott Guardian permission to review patient notes in time for the visit.

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## Haemophilia Comprehensive Care Centre

| Ref | Standard | Met? | Comments |
|---|---|---|---|
| HP-101 | **Service Information**<br><br>Written information should be offered to patients and, where appropriate, their carers covering at least:<br>a. Brief description of the service<br>b. Clinic times and how to change an appointment<br>c. Ward usually admitted to and its visiting times<br>d. Staff of the service<br>e. Community services and their contact numbers<br>f. Relevant national organisations and local support groups<br>g. Where to go in an emergency<br>h. How to:<br>i. Contact the service for help and advice, including out of hours<br>ii. Access social services<br>iii. Access benefits and immigration advice<br>iv. Interpreter and advocacy services, PALS, spiritual support and HealthWatch (or equivalent)<br>v. Give feedback on the service, including how to make a complaint<br>vi. Get involved in improving services (QS HP-199) | Y | However, reviewers did not see any evidence that this information was visible to patients visiting the Haemophilia Centre. |

| Ref | Standard | Met? | Comments |
|---|---|---|---|
| HP-102 | **Condition-Specific Information**<br><br>Written information should be offered to patients and, where appropriate, their carers covering at least:<br>a. A description of their condition and how it might affect them<br>b. How their condition is diagnosed<br>c. Genetics of inherited bleeding disorders<br>d. Testing for carrier status and the implications of being a carrier<br>e. Problems, symptoms and signs for which emergency advice should be sought<br>f. Out of hours services<br>g. 'On demand' clotting factor treatment<br>h. Prophylaxis<br>i. Self infusion (or infusion by parent or carer)<br>j. Home therapy and use of Haemtrack<br>k. How to manage bleeding at home<br>l. Ports, fistulae and in-dwelling access devices (if applicable)<br>m. Possible complications, including inhibitors and long term joint damage<br>n. Approach to elective and emergency surgery<br>o. Women's health issues<br>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<br>q. Dental care<br>r. Travel advice<br>s. Vaccination advice<br>t. National Haemophilia Database, its purpose and benefits<br>u. Sources of further advice and information<br><br>Information should be available covering:<br>1. Haemophilia A<br>2. Haemophilia B<br>3. Von Willebrand Disease<br>4. Acquired haemophilia<br>5. Inherited platelet disorders<br>6. Other less common and rare bleeding disorders | Y | However, reviewers did not see any evidence that this information was visible to patients visiting the Haemophilia Centre. All information provided was national or from drug companies. Information was not localised. |

| Ref | Standard | Met? | Comments |
|---|---|---|---|
| HP-103 | **Plan of Care**<br><br>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:<br>a. Agreed goals, including life-style goals<br>b. Self-management<br>c. Planned assessments, therapeutic and/or rehabilitation interventions<br>d. Early warning signs of problems, including acute exacerbations, and what to do if these occur<br>e. Agreed arrangements with school or other education provider and preparation for adult life (children and young people only)<br>f. Planned review date and how to access a review more quickly, if necessary<br>g. Who to contact with queries or for advice<br><br>The Plan of Care should be communicated to the patient's GP and to relevant other services involved in their care. | Not Assessed | Caldicott Guardian permission was not received from the Trust in time for the visit. This standard could not, therefore, be assessed by reviewers. |
| HP-104 | **Review of Plan of Care**<br><br>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. | Not Assessed | Caldicott Guardian permission was not received from the Trust in time for the visit. This standard could not, therefore, be assessed by reviewers. |
| HP-105 | **Contact for Queries and Advice**<br><br>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)**<br><br>All patients on home treatment should be encouraged to use electronic recording of their treatment through Haemtrack. | Y | However, compliance was noted to be low and there was no evidence of proactive follow-up by the haemophilia team of data submitted by patients. |

| Ref | Standard | Met? | Comments |
|---|---|---|---|
| HP-194 | **Environment**<br><br>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.<br><br>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. | N | See Concerns section of main report. |
| HP-195 | **Transition to Adult Services and Preparation for Adult Life**<br><br>Young people approaching the time when their care will transfer to adult services should be offered:<br>a. Information and support on taking responsibility for their own care<br>b. The opportunity to discuss the transfer of care with paediatric and adult services<br>c. A named coordinator for the transfer of care<br>d. A preparation period prior to transfer<br>e. Written information about the transfer of care including arrangements for monitoring during the time immediately afterwards<br>f. Advice for young people going away from home to study, including:<br>i. registering with a GP<br>ii. how to access emergency and routine care<br>iii. how to access support from their Comprehensive Care Centre<br>iv. communication with their new GP | Y | |
| HP-198 | **Carers' Needs**<br><br>Carers should be offered information on:<br>a. How to access an assessment of their own needs<br>b. What to do in an emergency<br>c. Services available to provide support | N | Reviewers did not see any evidence of documentation that encouraged carers to access information and support regarding their own emotional and wellbeing needs. |
| HP-199 | **Involving Patients and Carers**<br><br>The service should have:<br>a. Mechanisms for receiving regular feedback from patients and carers about treatment and care they receive<br>b. Mechanisms for involving patients and carers in decisions about the organisation of the service<br>c. Examples of changes made as a result of feedback and involvement of patients and carers | Y | However, the number of patient feedback returns was small, and evidence of changes made to the service as a result of feedback was limited (e.g. change to paediatric physiotherapy service). |

| Ref | Standard | Met? | Comments |
|---|---|---|---|
| HP-201 | **Lead Consultant and Lead Nurse**<br><br>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**<br><br>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:<br>a. Medical staff:<br>i. Consultant specialising in the care of people with inherited and acquired bleeding disorders available during normal working hours<br>ii. On-call consultant haematologist (24/7)<br>iii. Arrangements for advice from a consultant specialising the care of people with inherited and acquired bleeding disorders (if not on call)<br>b. Specialist nursing staff:<br>i. Bleeding disorders specialist nurses (5/7)<br>ii. Ward, out-patient and day unit staff with competences in the care of people with inherited and acquired bleeding disorders.<br>c. Clinical specialist physiotherapist<br>d. Biomedical Scientist and Clinical Scientist (further detail on the requirements are included in HP-303)<br>e. Clinical or counselling specialist psychologist or appropriately trained psychotherapist<br>f. Specialist senior social worker<br>g. Data manager | N | See the Concerns section of the main report. |

| Ref | Standard | Met? | Comments |
|---|---|---|---|
| HP-203 | **Service Competences and Training Plan**<br>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 | There was no evidence that the requirements of this standard in relation to a matrix identifying all members of the core team, their training requirements and their training achievements were being met. |
| HP-204 | **Competences – All Health and Social Care Professionals**<br>All health and social care professionals working in the service should have competences appropriate to their role in:<br>a. Safeguarding children and/or vulnerable adults<br>b. Recognising and meeting the needs of vulnerable children and/or adults<br>c. Dealing with challenging behaviour, violence and aggression<br>d. Mental Capacity Act and Deprivation of Liberty Safeguards<br>e. Resuscitation | Y | |
| HP-299 | **Administrative, Clerical and Data Collection Support**<br>Administrative, clerical and data collection support should be available. | Y | |
| HP-301 | **Support Services**<br>Timely access to the following support services should be available:<br>a. Play support (children's services only) including:<br>i. Play and distraction during any painful or invasive procedures<br>ii. Play support to enable the child's development and well-being<br>b. Pharmacy<br>c. Dietetics<br>d. Occupational Therapy<br>e. Orthotics | Y | |
| HP-302 | **Emergency Department – Staff Competences**<br>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:<br>a. Guidelines on care of patients with inherited and acquired bleeding disorders in the Emergency Department (QS HP-504)<br>b. Who to contact for advice | Y | However, patients who met the review team reported a lack of understanding from ED staff regarding bleeding disorders and an unwillingness of ED staff to follow the information contained on patients' bleeding disorder cards. |

| Ref | Standard | Met? | Comments |
|---|---|---|---|
| HP-303 | **Laboratory Service**<br><br>a. A UKAS / CPA accredited laboratory service with satisfactory External Quality Assurance performance should be available 24/7<br>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<br>c. The following tests should be available:<br>i. All coagulation factor assays (24/7)<br>ii. Inhibitor screening<br>iii. FVIII inhibitor quantification<br>iv. VWF antigen<br>v. VWF activity<br>vi. Platelet function testing<br>d. Molecular Genetic Laboratory service for:<br>i. detection of causative mutations in patients with inherited bleeding disorders<br>ii. carrier detection | Y | |
| HP-304 | **Specialist Services**<br><br>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:<br>a. Obstetrics including reproductive counselling, information about pre-implantation genetic diagnosis and antenatal diagnosis<br>b. Foetal medicine<br>c. Vascular access (consultant surgeon or interventional radiologist with experience of venous access devices)<br>d. Orthopaedic surgery<br>e. Care of older people services<br>f. Dental services<br>g. HIV services<br>h. Hepatology<br>i. Medical genetics (Genetic Counselling Services)<br>j. Pain management services<br>k. Rheumatology<br><br>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**<br><br>Facilities and equipment appropriate for the service provided should be available including:<br>a. Fridges<br>b. Storage<br>c. Clinical rooms for staff of all disciplines to see patients and carers<br>d. Room for multi-disciplinary discussion<br>e. Room for educational work with patients and carers<br>f. Office space for staff<br>g. Access to Haemtrack and the Haemophilia Centre Information System (HCIS) in all clinical areas<br><br>All equipment should be appropriately checked and maintained. | Y | |
| HP-499 | **IT System**<br><br>IT systems should be in use for:<br>a. Storage, retrieval and transmission of patient information, including access to the latest vCJD status and family tree<br>b. Patient administration, clinical records and outcome information<br>c. Data to support service improvement, audit and revalidation<br>d. Alerting the specialist team when patients attend the Emergency Department | Not assessed | Reviewers were unable to assess compliance with this standard as the Centre had not ensured that permission to review patients' records was received from the Trust's Caldicott Guardian in time for the review. |
| HP-501 | **Diagnosis Guidelines for Patients with Suspected Inherited and Acquired Bleeding Disorders**<br><br>Guidelines on diagnosis should be in use covering at least<br>a. Haemophilia A<br>b. Haemophilia B<br>c. Von Willebrand Disease<br>d. Acquired haemophilia<br>e. Inherited platelet disorders<br>f. Other less common and rare bleeding disorders | Y | |

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

| Ref | Standard | Met? | Comments |
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| HP-505 | **Guidelines on Care of Patients requiring Surgery**<br><br>Guidelines on the care of patients with inherited and acquired bleeding disorders who require surgery should be in use covering at least:<br>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<br>b. Communication of the agreed plan of care to all staff involved in the patient's care prior to, during and post-surgery<br>c. Documentation of care provided<br>d. Arrangements for escalation in the event of unexpected problems | Y | |
| HP-595 | **Guidelines on Transition and Preparing for Adult Life**<br><br>Guidelines on transition of young people from paediatric to adult services should be in use covering at least:<br>a. Taking responsibility for their own care<br>b. Involvement of the young person and, where appropriate, their carer in planning the transfer of care<br>c. Joint meeting between paediatric and adult services in order to plan the transfer<br>d. Allocation of a named coordinator for the transfer of care<br>e. A preparation period prior to transfer<br>f. Arrangements for monitoring during the time immediately after transfer<br>g. Advice for young people going away from home to study, including:<br>i. registering with a GP<br>ii. how to access emergency and routine care<br>iii. how to access support from their Comprehensive Care Centre<br>iv. communication with the young person's new GP | Y | See Good Practice section of main report. Patient feedback on transition was positive. |
| HP-599 | **Care of Vulnerable People**<br><br>Guidelines for the care of vulnerable children, young people and adults should be in use including:<br>a. Restraint and sedation<br>b. Missing patients<br>c. Mental Capacity Act and the Deprivation of Liberty Safeguards<br>d. Safeguarding<br>e. Information sharing<br>f. Palliative care<br>g. End of life care | Y | However, reviewers could not see any evidence for (b) – missing patients. |

| Ref | Standard | Met? | Comments |
|---|---|---|---|
| HP-601 | **Service Organisation**<br><br>The service should have an operational procedure covering at least:<br>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<br>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<br>c. Responsibility for giving information and education at each stage of the patient journey<br>d. Arrangements for involving Haemophilia Centre staff in multi-disciplinary discussions relating to their patients (QS HP-602)<br>e. Arrangements for follow up of patients who 'do not attend'<br>f. Arrangements for transfer of patient information when patients move areas temporarily or permanently<br>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)<br>h. Ensuring school visits for children with severe haemophilia at least at each change of school (children's services only)<br>i. Ensuring patients are visited at home at least annually if they are unable to attend clinics, including those in nursing homes<br>j. Lone working | N | Reviewers did not see any evidence of an operational policy for the service. |
| HP-602 | **Multi-Disciplinary Team Meetings**<br><br>Multi-disciplinary team meetings to discuss patients' plans of care should take place regularly involving:<br>a. All core members of the specialist team (HP-202)<br>b. Senior biomedical scientist or clinical scientist with responsibility for the Coagulation Laboratory<br>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**<br><br>The following multi-disciplinary clinic arrangements for patients with inherited and acquired bleeding disorders should be in place:<br>a. Involvement of medical, specialist nursing and physiotherapy staff in clinics<br>b. Availability of social work and psychology staff in clinics<br>c. Combined clinics or other arrangements for multi-disciplinary discussion with:<br>i. orthopaedics<br>ii. rheumatology<br>iii. obstetrics and gynaecology<br>iv. paediatrics<br>v. dental<br>vi. HIV / hepatology | N | Reviewers did not see any evidence of regular clinics involving all team members identified in (a). In addition, there was no input from psychology or social work staff. |
| HP-604 | **Liaison with Other Services**<br><br>Review meetings should be held at least annually with specialist services to consider liaison arrangements and address any problems identified. | N | Reviewers did not see any evidence of regular formal meetings with colleagues from other relevant specialist services. |
| HP-701 | **Data Collection**<br><br>The following data should be collected:<br>a. UK National Haemophilia Database data on all patients<br>b. Data on concentrate use and bleeds, either through Haemtrack or an equivalent mechanism<br>c. Data required to complete the UKHCDO National Haemophilia Dashboard or other national mechanisms | N | Although data was being submitted, there was a lack of clarity regarding which patients were the sole responsibility of the Southampton team. Different sets figures were presented before and after the visit. See Concerns section of main report. |
| HP-702 | **Audit**<br><br>The services should have a rolling programme of audit covering at least:<br>a. Clinical guidelines (QS HP-503)<br>b. Emergency and out of hours care (QS HP-504)<br>c. Initiation of prophylaxis in children<br>d. Inhibitor surveillance and Immune Tolerance Induction (ITI)<br>e. Clinical reviews including joint scores (QS HP-103 & 104)<br>f. Concentrate use and wastage | N | Reviewers did see some evidence of audit activity, but this activity did not meet all the requirements identified for this standard and there was no documented rolling audit programme in place. |
| HP-706 | **Research**<br><br>The service should actively participate in research relating to the care of patients with bleeding disorders. | Y | See Achievements section of main report. |

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

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## Network

| Ref | Standard | Met? | Comments |
|---|---|---|---|
| HY-199 | **Involving Patients and Carers**<br><br>The network should have mechanisms for involving patients and their carers from all services in the work of the network. | N | Reviewers did not see any evidence of mechanisms for involving patients and receiving their feedback across the network. |
| HY-203 | **Inherited and Acquired Bleeding Disorders Network Leads**<br><br>The network should have a nominated:<br>a. Lead consultant and deputy<br>b. Lead specialist nurse<br>c. Lead physiotherapist<br>d. Lead clinical or counselling psychologist<br>e. Lead manager | N | Reviewers did not see any evidence of leadership or participation in a formal network arrangement. |
| HY-204 | **Education and Training**<br><br>The network should have agreed a programme of education and training to help services achieve compliance with QSs HP-203. | N | Although there was some evidence of education and training between nurse colleagues, reviewers did not see any evidence of this being part of a formal network arrangement. |
| HY-503 | **Guidelines**<br><br>Network guidelines should have been agreed covering:<br>a. Diagnosis for patients with suspected inherited and acquired bleeding disorders (QS HP-501)<br>b. Concentrate use and monitoring (QS HP-502)<br>c. Clinical guidelines (QS HP-503)<br>d. Management of patients with inherited and acquired bleeding disorders in the Emergency Department (QS HP-504)<br>e. Care of patients requiring surgery (QS HP-505)<br>f. Transition and preparing for adult life (QS HP-595) | N | Reviewers did not see any evidence of mechanisms for sharing guidelines across the network. |
| HY-701 | **Ongoing Monitoring**<br><br>The network should monitor on a regular basis:<br>a. Submission of data on all patients to the UK National Haemophilia Database (QS HP-701)<br>b. Network-wide data on concentrate use and bleeds | N | Reviewers did not see any evidence of mechanisms for the ongoing monitoring and sharing of data across the network. |
| HY-702 | **Audit**<br><br>The network should have an agreed programme of audit and review covering network-wide achievement of QS HP-702. | N | Reviewers did not see any evidence of mechanisms for audit across the network. |
| HY-703 | **Research**<br><br>The network should have agreed:<br>a. A policy on access to research relating to the care of patients with inherited and acquired bleeding disorders<br>b. A list of research trials available to all patients within the network. | N | Reviewers did not see any evidence of a network research policy. |

| Ref | Standard | Met? | Comments |
|---|---|---|---|
| HY-798 | **Network Review and Learning**<br><br>Representatives of Comprehensive Care Centres and referring Haemophilia Centres should meet at least once a year to:<br>a. Identify any changes needed to network-wide policies, procedures and guidelines<br>b. Review results of audits undertaken and agree action plans<br>c. Review and agree learning from positive feedback, complaints, critical incidents and 'near misses', including those involving liaison between teams<br>d. Share good practice and potential service improvements | N | Reviewers did not see any evidence network review and learning. |

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## Commissioning

| Ref | Standard | Met? | Comments |
|---|---|---|---|
| HZ-601 | **Commissioning of Services**<br><br>Commissioners should have agreed the configuration of clinical networks including:<br>a. Designated Comprehensive Care Centres and Haemophilia Centres and the relationships between them<br>b. Whether the service cares for children, adults or both<br>c. Referral pattern to each service, taking into account the type of patients who will be treated by each team | N | Reviewers were concerned that agreed patient pathways for paediatrics were not fully understood or agreed between commissioners and providers |
| HZ-701 | **Clinical Quality Review Meetings**<br><br>Commissioners should regularly review the quality of care provided by:<br>a. Each service, including achievement of QS HP-701<br>b. Each network, including achievement of QS HY-701 and QS HY-798<br>c. Service and network achievement of relevant QSs | Y | Meetings were held at least twice a year to review the service, and to review factor consumption and planning for the coming year. In addition, a Wessex wide meeting was held at least twice a year to review the whole region and members of the Southampton team attended these meetings. |
| HZ-798 | **Network Review and Learning**<br><br>Commissioners should attend a Network Review and Learning meeting (HY-798) at least once a year for each network in their area. | Y | See HZ-701 |

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