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Specialised Services Service Specification: Inherited Bleeding Disorders

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Approved by:	Management Group
Issue Date:	19 May 2014
Review Date:	May 2017
Document No:	CP77

Document History

Revision History			
Version No.	Revision date	Summary of Changes	Updated to version no.:
		New document based on English service specification	0.1
0.1	19/05/2014	Ratified	1.0
Date of next revision			

Consultation		
Name	Date of Issue	Version Number
Inherited Bleeding Disorders Advisory Group	30/01/2014	0.1
Cancer Programme Team	24/04/2014	0.1
Royal Liverpool and Broadgreen NHS Trust	17/04/2014	0.1

Approvals		
Name	Date of Issue	Version No.
Executive Directors	25/04/2014	0.1
Management Group	08/05/2014	0.1

Distribution – this document has been distributed to			
Name	By	Date of Issue	Version No.
WHSSC Website	Corporate	19/05/2014	1.0
LHB Medical Directors	Corporate	19/05/2014	1.0
Providers	Planning	19/05/2014	1.0

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1. Aim

1.1 Introduction

The document has been developed as the service specification for the planning of inherited bleeding disorders for patients resident in Wales.

The purpose of this document is to:

- Detail the specification for the inherited bleeding disorder services for patients that are included in the Welsh population for commissioning purposes.

Specifically, all adults and children with the following diagnoses will have access to the service:

- Haemophilia A (Factor VIII deficiency)
- Haemophilia B (Factor IX deficiency)
- Von Willebrand Disease
- Acquired Haemophilia and other related bleeding disorders
- Other rare forms of Inherited Bleeding Disorders

A full list of ICD10 codes are included in the Commissioning Policy.

There is a spectrum of severity for each of these conditions; however there is no national definition of when a 'mild' bleeding disorder requires specialist intervention. The service should therefore cover all severities.

Specialist services for haemophilia and other bleeding disorders specifically do not cover the following:

- The use of clotting factor for treatment of trauma or critical care or following surgery or as part of obstetric care (where the patient has no underlying bleeding disorder)
- General haemostasis and thrombosis services (including stroke services)
- Specialist haemostasis and thrombosis unrelated to haemophilia and bleeding disorders (e.g. liver disease).
- Anticoagulation treatment or the reversal of anticoagulant treatment.

1.2 Relationship with other Policies and Service Specifications

This document should be read in conjunction with the following documents:

- Commissioning Policy for Inherited Bleeding Disorders.

2. Service Delivery

2.1 Service Model/Pathway

Initial referral will be due to a suspected or known bleeding disorder and could come from a GP or as a referral to tertiary care from an acute Trust. Either route could follow clinical presentation, laboratory investigations showing abnormal coagulation or could be due to genetic diagnosis and/or family studies.

The standard service model for haemophilia services was set out in the 'English Health Service Guidance (HSG) (93) 30 Provision of Haemophilia Treatment and Care.' This states that there should be two different levels of haemophilia provision - a Comprehensive Care Centre (CCC) which provides specialist diagnosis and care and a Haemophilia Centre (HC) which provides a local, shared care service. This Service Specification aims to further develop and strengthen this model of care.

Haemophilia Centres must be part of a comprehensive care programme network with a Comprehensive Care Centre. This must be formally agreed and documented.

The providers of services for Welsh patients are:

- Comprehensive Care Centre, University Hospital of Wales, C&V UHB
- Haemophilia Centre, Singleton Hospital, ABM UHB
- Comprehensive Care Centre, Royal Liverpool and Broadgreen NHS Trust
- Haemophilia Centre, Ysbyty Gwynedd, BC UHB
- Comprehensive Care Centre, Alder Hey Children's NHS Foundation Trust.

The model of care will ensure patients have access to comprehensive care which includes the following:

- A clinical service provided by healthcare professionals experienced in the treatment of haemophilia and other bleeding disorders, including Consultant Haematologists with a specialist interest in haemostasis.
- Specialist Nurses trained in line with the Haemophilia Nurses Association and Specialist Physiotherapists trained in line with the Haemophilia Chartered Physiotherapy Association.
- 24 hour laboratory service accredited by United Kingdom Accreditation Service (UKAS) capable of carrying out all tests necessary for the definitive diagnosis of haemophilia and other bleeding disorders, including the identification and assay of the relevant specific haemostatic factors. These laboratories will be capable of monitoring therapy and screening for inhibitors with quantification of any inhibitor detected in accordance with published UKHCDO guidelines.
- Maintenance of satisfactory quality control and assurance for all laboratory tests offered in relation to clinical services, both by establishing the appropriate level in the UK National External Quality Assessment Scheme in blood coagulation (NEQAS), or other relevant approved external quality assessment schemes.
- Further investigation of relatives of patients with haemophilia and other bleeding disorders. The Comprehensive Care Centre service will also include the diagnosis of atypical cases, genotypic analysis, the assay of inhibitors, haemostatic factors, diagnosis of hereditary platelet disorders and molecular diagnostic testing. Haemophilia Centres should refer to the CCC for the specialist elements of care where they are not available locally.
- An advisory service to patients and close relatives on matters specific to haemophilia and other bleeding disorders. Advice will also be provided to other healthcare professionals. A Comprehensive Care Centre (CCC) will provide 24-hour advisory service to Haemophilia Centres within their managed clinical network and support to such centres as appropriate.
- Musculoskeletal services to include access to and regular review by experienced specialist physiotherapists.
- Paediatric services to be provided by personnel trained in the care of children.
- When treating children, the service will additionally follow current standards and guidance for the safe treatment of children.
- Access and referral to clinical services for all aspects of haemophilia care including:
 - Orthopaedic surgical services
 - Dental services
 - HIV and hepatology services including access to a Consultant in Hepatology.

- Counselling services, preferably to a named counsellor, who has experience of patients with haemophilia and inherited bleeding disorders
- Obstetric services, including reproductive counselling.
- Social care support.
- Good quality medical records will be maintained for all patients. Records must be maintained of all treatment administered and all adverse reactions reported. All relevant patients will be formally registered with the treating Centre and with the UKHCDO National Haemophilia Database.
- Special medical cards will be issued which include details of the patient's bleeding disorder, usual treatment, treatment centre and out of hours contact details for the centre.
- Education facilities and training opportunities for medical staff, nurses, counsellors and other personnel as required, promoting optimal comprehensive care of patients.
- Undertaking research programmes, including the conduct of clinical trials and to establish and participate in suitable regional and national programmes of clinical audit.

A home therapy programme should support home treatment for relevant patients across the network including the administration of prophylactic therapy and home and school visits where appropriate.

Providers will purchase clotting factor products via the national framework agreement.

Transfer planning will take place between the paediatric and adult provider to ensure a smooth transition to adult care for adolescents.

Transfer planning should take place to a paediatric provider for the management of neonates with antenatal diagnosis of, or known to be at risk of, haemophilia and other bleeding disorders.

Pregnancy

Pregnant women with pre-existing conditions as discussed in this specification require assessment and/or management from specialist tertiary maternity care delivered within a dedicated multidisciplinary service staffed by a maternal medicine specialist, a physician, and supporting multidisciplinary team with extensive experience of managing the condition in pregnancy.

In view of this, local haemophilia centres must have formal outreach arrangements and transfer protocols in place with specialised tertiary

maternity units with access to appropriate tertiary medical, surgical, fetal medicine, clinical genetics and level 3 Neonatal Intensive Care services. These specialised maternity services must have a critical mass of activity to maintain expertise, ensure best practice, training opportunities and for the organisational infrastructure, staffing, facilities and equipment to be clinically and economically efficient. They should have robust risk management and performance monitoring processes.

All such women must receive personalised pre-pregnancy and maternity care planning from specialised tertiary maternity services to allow optimal disease management in the context of the pregnancy. This will reduce avoidable morbidity, mortality and unnecessary intervention for mother and baby.

Women with conditions as discussed in this specification must be referred immediately once they are pregnant to plan their care. This must include access to termination of pregnancy and specialist advice regarding contraception. The individualised care plan must cover the ante natal, intrapartum and postnatal periods. It must include clear instructions for shared care with secondary services, when appropriate including escalation and transfer protocols and clear guidelines for planned and emergency delivery.

Interdependencies with other services

In addition to the specialised elements of care there are also a number of general services with varying levels of interdependency with haemophilia and other bleeding disorders.

Co-located services

Comprehensive Care Centres will have the following on-site services:

- 24 hour laboratory service capable of carrying out all tests necessary for the definitive diagnosis of haemophilia and other bleeding disorders

Interdependent services

Services that are required during the spell of care but where there is no absolute requirement for this service to be based on the same healthcare delivery site are as follows:

- Dental services.
- HIV and hepatology services including access to Consultant Hepatology input.

- Counselling services, preferably to a named counsellor, who has experience of patients with haemophilia and inherited bleeding disorders.
- Obstetric services, including reproductive counselling.
- Social care support.

Related services

There are a number of specialised services with a clear inter-dependency for specific patient sub-groups of those with haemophilia and other bleeding disorders as follows:

- Specialised services for women's health – this is relevant for maternity services and fetal medicine for women who are carriers or have a bleeding disorder
- Specialised services for liver, biliary and pancreatic medicine and surgery (adult) – this relates to access to hepatology for hepatitis-infected patients.
- Medical genetic services.
- Specialised services for children – important inclusions are anaesthesia and pain management, dentistry, NIC, PIC and paediatric surgery.
- Specialised rheumatology services (all ages).
- Specialised pain management services (adult).
- Specialised orthopaedic services (adult).

Additionally, for the group of infected individuals there is a need for further inter-dependency with the following services:

- HIV/AIDS Treatment and Care and associated services

The service model needs to consider access arrangements for other specialised services, and the funding and quality assurance responsibilities lie with Health Boards where they are not commissioned by WHSSC.

2.2 Population Covered

The population covered is the whole of Wales subject to the current guidance regarding the border population. For the purpose of commissioning health services this includes patients who are resident in England but who are registered with a GP practice in Wales.

3. Quality and Patient Safety

3.1 Quality and Patient Safety

The providers must work to written quality standards and provide immediate information to WHSSC regarding serious clinical incidents, serious or escalating numbers of complaints and/or issues which may entail media or political interest.

Services must be accessible to all patients with an inherited bleeding disorder regardless of sex, race or gender. There will be clear and agreed pathways within the managed clinical network to ensure that all patients have access to comprehensive care. The patient pathway must ensure that all patients have access to comprehensive care 24 hours a day, 7 days a week, including protocols for out-of-hours care, emergency management and the treatment of inhibitors. Staff should attend mandatory training on equality and diversity and the facilities provided should offer appropriate disabled access for patients, family and carers. When required, the providers will use translators and printed information available in multiple languages, including Welsh.

3.2 Quality Indicators and Standards

The clinical service will be provided by healthcare professionals experienced in the treatment of haemophilia and other bleeding disorders. This includes Haematology Consultants with a specialist interest in Haemostasis, Specialist Nurses trained in line with Haemophilia Nurses Association and Specialist Physiotherapists trained in line with the Haemophilia Chartered Physiotherapy Association.

All CCCs must participate in the UKHCDO triennial audit programme and achieve the accreditation standards. CCCs will normally provide treatment for 40 or more severely affected patients per year.

There will be clear and agreed pathways within the comprehensive care programme to ensure that all patients have access to comprehensive care. The patient pathway must ensure that all patients have access to comprehensive care 24 hours a day, 7 days a week, including protocols for out-of-hours care, emergency management and the treatment of inhibitors.

Patient reviews will take place in accordance with this service specification within a multidisciplinary team. As a minimum all patients with haemophilia and other bleeding disorders must have contact with their CCC at least once a year, for those patients not receiving treatment this may be a structured telephone interview. Severe patients must have 6 monthly reviews and young children (<5yrs) must be seen 3-4 times per year.

Providers must also ensure complete, accurate and timely returns to the National Haemophilia Database. The annual report of the UKHCDO data will be formally reviewed by the Commissioners and the All-Wales IBD Advisory Group. The UKHCDO triennial audit report will also be used to ensure quality of service for Welsh patients.

The following national documents are also of relevance to this service specification:

- Model Documentation for Designation of Haemophilia Services, London
- Specialised Commissioning Group (June 2010)
- United Kingdom Haemophilia Centre Directors Organisation (UKHCDO) Triennial
- Audit Report (2011)
- UKHCDO Clinical Outcomes Group Report (2011)
- All United Kingdom Haemophilia Centre Doctors Organisation (UKHCDO) Guidelines
- A National Service Specification for Haemophilia and Related conditions (The Haemophilia Alliance), 2006.

3.3 Putting Things Right: Raising a Concern

Whilst every effort has been made to ensure that decisions made under this policy are robust and appropriate for the patient group, it is acknowledged that there may be occasions when the patient or their representative are not happy with decisions made or the treatment provided. The patient or their representative should be guided by the clinician, or the member of NHS staff with whom the concern is raised, to the appropriate arrangements for management of their concern:

- When a patient or their representative is unhappy with the decision that the patient does not meet the criteria for treatment further information can be provided demonstrating exceptionality. The request will then be considered by the All Wales IPFR Panel;
- If the patient or their representative is not happy with the decision of the All Wales IPFR Panel the patient and/or their representative has a right to ask for this decision to be reviewed. The grounds for the review, which are detailed in the All Wales Policy: Making Decisions on Individual Patient Funding Requests (IPFR), must be clearly stated. The review should be undertaken, by the patient's Local Health Board;
- When a patient or their representative is unhappy with the care provided during the treatment or the clinical decision to withdraw treatment provided under this policy, the patient and/or their representative should be guided to the LHB for NHS Putting Things Right. For services provided outside NHS Wales the patient or their representative should be guided to the NHS Trust Concerns Procedure with a copy of the concern being sent to WHSSC.

4. Performance Monitoring and Information Requirements

4.1 Performance Monitoring

WHSSC will be responsible for commissioning services in line with this policy. This will include agreeing appropriate information and procedures to monitor the performance of organisations.

For the services defined in this policy the following approach will be adopted:

- **Service providers to evidence quality and performance controls**
- **Service providers to evidence compliance with standards of care**

WHSSC will conduct performance and quality reviews on an annual basis.

4.2 Key Performance Indicators

The providers will be expected to monitor outcomes by complying with the UKHCDO data requirements for all patients.

The provider should also monitor the appropriateness of referrals into the service and provide regular feedback to referrers on inappropriate referrals, identifying any trends or potential educational needs.

5. Equality Impact and Assessment

The Equality Impact Assessment (EQIA) process has been developed to help promote fair and equal treatment in the delivery of health services. It aims to enable Welsh Health Specialised Services Committee to identify and eliminate detrimental treatment caused by the adverse impact of health service policies upon groups and individuals for reasons of race, gender re-assignment, disability, sex, sexual orientation, age, religion and belief, marriage and civil partnership, pregnancy and maternity and language (Welsh).

This policy has been subjected to an Equality Impact Assessment.

The Assessment demonstrates the policy is robust and there is no potential for discrimination or adverse impact. All opportunities to promote equality have been taken.