CLINICAL GUIDELINE FOR THE MANAGEMENT OF PRIMARY HAEMOSTATIC DISORDERS (Haemophilia & Related Conditions)

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1. **Aim/Purpose of this Guideline**

This document outlines the basis of care for the management of patients with disorders of haemostasis, by the Royal Cornwall Hospitals NHS Trust Haemophilia Centre (Department of Haematology RCH-T), in accordance with current recommendations and the National Service Specification for Haemophilia and Related Conditions (NHS England 2013).

2. **The Guidance**

2.1. **Glossary**

**APTT** - The activated partial thromboplastin time.

**Desmopressin (acetate)** – (brand name Octim®) synthetic anti-diuretic hormone administered IV, SC or intra-nasally. Has the physiological effect of increasing factor VIII and Von Willebrand Factor levels.

**Factor** - When followed by a Roman numeral e.g. Factor VIII or Factor IX, this describes one of the proteins of the body's clotting pathway, expressed as international units per dl (IU/dl). When such a nomenclature is followed by a level this describes the laboratory estimation of the patient’s protein with reference to a normal range.

**FEIBA concentrate** – Factor VIII Inhibitor Bypassing Fraction, containing an anti-FVIII inhibitor coagulant complex of standardised activity, with factor VIII coagulant antigen (FVIII C:Ag) present in a concentration of up to 0.1 U/1 U FEIBA.

**Haemarthrosis** - A bleed into a joint.

**Haemophilia** - This describes the clinical condition associated with either Factor VIII deficiency, i.e. haemophilia A or Factor IX deficiency i.e. haemophilia B (also called Christmas disease).

**Haemostasis** - The natural system for formation of blood clots by a process of activation of blood factors together with platelet cells.

**Inhibitor studies** - Mixing studies with normal plasma to determine if there is correction of the abnormal screening test, followed by specific factor inhibitor assays.

**Muco-cutaneous** - A term used to describe bleeding of mucoid surfaces such as the mouth, nasopharangeal passages, or endometrium, and/or the skin.

**Plasma** - The fluid component of blood, containing clotting factors in solution.

**PT** - Prothrombin time.

**Recombinant** - Descriptive term for a product produced by genetic engineering, for example FVIII (r-FVIII) or recombinant activated factor VII (r-FVIIa).
2.2. Symptoms and the Investigation of Primary Haemostatic Disorders

Bruising  A common phenomenon, which is often non-discriminatory between normal persons and patients with disorders of haemostasis. Common in Von Willebrand’s disease and platelet disorders.

Joint bleeds  Usually occurs in patients with isolated factor deficiencies, such as haemophilia A and B, causing acute pain often with associated swelling and warmth. It may be isolated to particular target joints, e.g. knee or ankle.

Muco-cutaneous bleeding  Eg epistaxes (nose bleeds) and menorrhagia (heavy periods)

Muscle bleeds  Present with pain, swelling and possible loss of use of the associated limb. Occurs with both isolated factor disorders and platelet defects. Potentially more troublesome than joint bleeds, as they are not limited by a capsule.

2.3. Investigation

Level 1 - Basic Screen
A “coagulation screen” of the platelet count, PT and APTT are basic initial tests.

Level 2
In patients with a history suspicious of a primary bleeding diathesis, von Willebrand’s disease is most commonly identified and screening tests should be:

Platelet count and morphology
Blood group
PT and APTT
Factor VIII complex (testing both the factor VIII clotting level together with the Von Willebrand factor by antigenic and functional activity assay) to be received in laboratory the same day
Bleeding time (or in children consider sample to Derriford for PFA-100 analysis)

(NB paediatric cases minimum sample for PT, APTT, FVIII complex is 2mls in paediatric tubes)

Consider platelet function tests in patients with a significant history (20ml sample) to be received in laboratory within 60 minutes. Ensure that requests for coagulation tests other than PT and APTT are discussed with laboratory staff before sampling the patient. Some tests require special setup in the laboratory.

Level 3
In patients with prolonged prothrombin time or APTT not otherwise explained, specific factor assays will be done with inhibitor studies. Patients with a normal coagulation screen but abnormal bleeding time should undergo platelet function tests and nucleotide assays, as indicated, arranged with the laboratory. Platelet Function tests require liaison with the laboratory before sampling the patient.
Level 4
Where a specific diagnosis is indicated supplementary tests may be required eg: Von Willebrand’s disease: RIPA, collagen binding assay, multimeric analysis
Haemophilia: genetic analysis

2.4. Clinical classification of bleeding disorders

2.4.1. Summary for Haemophilia A & Haemophilia B

<table>
<thead>
<tr>
<th>Phenotype</th>
<th>Factor Level IU/dl</th>
<th>Clinical Bleeding (typically of the joints)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>&gt;5</td>
<td>Infrequent usually resulting from trauma.</td>
</tr>
<tr>
<td>Moderate</td>
<td>1 – 5</td>
<td>Infrequent, occasionally spontaneous.</td>
</tr>
<tr>
<td>Severe</td>
<td>&lt;1</td>
<td>Frequent and spontaneous</td>
</tr>
</tbody>
</table>

2.4.2. Von Willebrand’s disease
This is different to haemophilia as the severity depends on quantitative and, or qualitative reductions in von Willebrand factor multimers. Bleeding is more commonly muco-cutaneous.

<table>
<thead>
<tr>
<th>Type</th>
<th>Von Willebrand factor multimers</th>
<th>disease severity</th>
<th>management</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>quantitative reduction</td>
<td>mild</td>
<td>usually desmopressin</td>
</tr>
<tr>
<td>2</td>
<td>subtyped e.g. 2a, 2b, 2bN by quantitative and qualitative reduction</td>
<td>mild</td>
<td>usually factor (derived from human plasma)</td>
</tr>
<tr>
<td>3</td>
<td>severe reduction of all multimers</td>
<td>severe</td>
<td>Factor (derived from human plasma)</td>
</tr>
</tbody>
</table>

2.4.3. Factor XI deficiency
This is relatively rare. Patients rarely bleed unless the factor level is <15 iu/dl and this is usually muco-cutaneous (ENT or GU).

2.4.4. Factor XII deficiency
Usually recognised as the cause for a prolonged APTT in otherwise healthy patients. Bleeding is most unusual and specific therapy does not appear to be necessary.

2.4.5. Rare deficiencies
Other single coagulation factor deficiencies affect eg factors II, V, VII, X, XI and XIII.

2.5. Patient registration

Departmental excel spreadsheet database
All patients registered with the RCH-T Haemophilia Centre are recorded on an excel spreadsheet (see section 2.11.3) with their diagnosis and treatment of choice. This database is also held in the transfusion Section of the Laboratory and issued to relevant clinical areas.
**Hospital notes**
At the time of registration a brief résumé of the diagnosis, blood levels, blood group and where appropriate the response to desmopressin should be recorded on the inside cover of the hospital notes.

The Hospital notes of patients with relatively frequent bleeding will usually be held on the Treliske site in the Medical Records Mega-shed, those with severe disorders in the Department of Haematology.

The patient's PAS & Oceano record will state ‘Ref Haemostasis Guidance’

**Red UKHCDO Bleeding Disorder Cards**
All patients are issued these cards from the Haematology Department, detailing the nature of their disorder, their Clinician and contact number. They are instructed to carry the card with them and to produce for attending clinical staff in the event of bleeding, or other challenges. If the patient is responsive to specific therapy such as desmopressin this will be marked on the card.

**United Kingdom Haemophilia Centres Doctor’s Organisation Database**
All patients should be registered after explanation of this national database with the patient’s, or parental, verbal consent, using the appropriate forms, or via secure internet connection. An information sheet is available. Notification of adverse events and new inhibitor patients is discussed below.

**2.6. Treatment Recommendations**

**2.6.1. On-Call**
All patients who may be bleeding may self-refer and have 24 hour access to emergency care as follows:

<table>
<thead>
<tr>
<th></th>
<th>Within working hours</th>
<th>outside working hours</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adults</td>
<td>contact and attend the Department</td>
<td>contact Lowen Ward, who will arrange for assessment in the Emergency Department</td>
</tr>
<tr>
<td>Children under 18 years</td>
<td>“ “ “</td>
<td>contact Paed Obs Unit</td>
</tr>
</tbody>
</table>

Patients with non-bleeding problems will be sent in by the usual means eg the out of hours service currently SERCO Health.

The On-Call Clinical Haematologist *should routinely be informed* of a patient’s attendance, either with or without specific bleeding problems.

**2.6.2. Acute Management of Bleeding**
This is dependent on the nature of the bleed, the deficiency and its severity, It should be instituted promptly and, Patients should never be discharged without treatment unless in consultation with the on-call haematology consultant,
Where possible refer to the patients details on their red Bleeding Disorders Card and the Haematology Department’s database, held in the Transfusion Laboratory with printed copies in acute clinical areas, which lists the choices for appropriate treatment:

**Investigations**
If a severe bleed check the full blood count and take a citrate (coagulation) sample for “freeze plasma”. Group and screen if indicated, eg haematemesis, There is nothing to be gained by repeating a patient’s APTT, or baseline level, as this will be prolonged as per usual.

**Haemophilia A treatment of choice**
This depends on the phenotypic severity, cf. above.

<table>
<thead>
<tr>
<th>Mild</th>
<th>Usually desmopressin (cf. below).</th>
</tr>
</thead>
<tbody>
<tr>
<td>Moderate</td>
<td>Usually factor VIII concentrate</td>
</tr>
<tr>
<td>Severe</td>
<td>Always factor VIII concentrate</td>
</tr>
<tr>
<td>Head injury</td>
<td>Factor concentrate, desmopressin is a relative contra-indication</td>
</tr>
</tbody>
</table>

**Haemophilia B**
These patients do not respond to desmopressin and require specific concentrate therapy

**Haemophilia A & B management outline:**

<table>
<thead>
<tr>
<th>Condition/bleed site</th>
<th>Requisite post-treatment factor level IU/dl for haemostasis</th>
<th>Additional Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>Minor spontaneous bleeding or joint bleed</td>
<td>20-40</td>
<td>Consider admission for bed rest or review following day</td>
</tr>
<tr>
<td>Severe bleeding, e.g. muscle, joint or profuse haematuria</td>
<td>30-60</td>
<td>Admit for bed rest</td>
</tr>
<tr>
<td>*Head injury</td>
<td>80-100</td>
<td><strong>Urgent CT. BD therapy</strong>, or by infusion - consider factor levels.</td>
</tr>
<tr>
<td>Major surgery</td>
<td>80-100</td>
<td>BD therapy**, or by infusion - consider factor levels.</td>
</tr>
</tbody>
</table>

*Significant head injury, as associated with loss of consciousness, confusion, nausea, deep wound, haematoma or fracture. **OD in haemophilia B
NB Ileopsoas muscle bleeds
Often the patient will complain of pain in the hip and will present holding the hip in flexion,
Tenderness may be minimal but extension of the hip is impossible,
A neurological deficit of the femoral region must be excluded as this may indicate acute compression of the femoral nerve with consequent morbidity,
These patients are always admitted and treated aggressively, with target factor VIIIc 80-100 IU/dl.

Von Willebrand’s disease
Most patients have mild type 1 disease and have an excellent response to desmopressin,
Desmopressin is best avoided in the treatment of significant head injury.
Type 2 patients have a variable response to desmopressin, whilst this ineffective in type 3 disease, when factor concentrate is required for control of haemorrhage.

Rare factor deficiencies
These may require factor concentrate, as below, or for milder bleeding tranexamic acid may be appropriate, as advised by the on-call haematology consultant.

2.6.3. Factor Concentrate Dosage see also appendix A
The dosage is roughly calculated using a series of formulae based on known factor half-life and kinetics
When bleeding is severe post-infusion levels may be taken to ensure adequate treatment, thereafter repeat levels are at defined intervals.

Haemophilia A (factor VIII deficiency)
Recombinant Factor VIII (eg, Advate, Re-Facto AF and Helixate NexGen) or
Intermediate purity Haemate P,
Dose either as target IU/kg, as per appendix A, or by target rise in IU/dl when
the dose = patient’s weight x (target IU/dl - baseline factor level) x 0.5

Haemophilia B (factor IX deficiency)
Recombinant Factor IX Benefix or
Dose either as target IU/kg, or by target rise rise in IU/dl when
the dose = patient’s weight x (target IU/dl - baseline factor level) x 1.4

Von Willebrand’s Disease
Intermediate human derived purity Factor VIII Concentrate eg Haemate P,
the dose = patient’s weight x (target IU/dl - baseline factor level) x 0.5

2.6.4. Other deficiencies for which concentrates are available

Factor XI Deficiency

Fresh frozen plasma (FFP) 15-20 ml/kg
or BPL Factor XI Concentrate
the dose = patient’s weight x (target IU/dl - baseline factor level) x 0.5
NB The post infusion level should be checked
The total dose of concentrate should not exceed 30 U/kg and peak factor XI 100 IU/dl,
otherwise there is a significant risk of thrombosis

2.6.5. Desmopressin acetate (OCTIM 15microgrammes/ml ampoule).
Different preparations are available – for subcutaneous use please order from pharmacy by brand name. Out of hours stock is available from the fridge in MAU, ED & Lowen Ward

- This synthetic analogue of anti-diuretic hormone will boost factor VIII and von Willebrand factor levels 2 to 4-fold by promoting endothelial cell release.
- It is given once daily and often the effect wanes after the first one or two injections.
- As it activates the fibrinolytic pathways, tranexamic acid will be given in conjunction.
- Due to its anti-diuretic effect, patients may be at risk of water intoxication with hyponatraemia and at worst convulsions and coma.
- Therefore patients should be water-restricted, drinking only to quench their thirst (eg adults use maximum 2 litres or for children 2/3rd of usual maintenance), for the following 24 hours and with repeated administration check U&E, weight and urine output daily.
- It is (generally) contra-indicated in children under two years of age.
- It may be administered by the SC, IV or intra-nasal routes

Patients will usually have undergone a trial of desmopressin to determine whether this is suitable therapy as reported on the database and their red Bleeding Disorder card.

Dose = 0.3 micrograms/kg either undiluted SC, or IV in 50-100 ml saline over 20 minutes.

Tranexamic acid – orally: 25 mg/kg tds for 7 days (adults typically 1 g tds)
I.V: 10mg/kg tds
Mouth wash: cf dental surgery section.

N.B. Tranexamic acid should not be used in patients with haematuria, pregnancy or those with a thrombotic history.

Desmopressin given subcutaneously (OCTIM 15microgrammes/ml ampoule) is equally efficacious to IV administration and probably causes fewer side effects. There is also an intra-nasal preparation (OCTIM nasal spray 150 micrograms per actuation), 4 micrograms/kg, usual adult dose 300 micrograms, ie two actuations (sprays) once every three days. This must be sprayed and not sniffed as this reduces absorption. The activity is equivalent to about 60% of the IV/SC preparation.

2.6.6. Platelets
In either quantitative or qualitative platelet disorders, transfusion of platelets may be required, although these are generally used sparingly as patients may develop antibodies. On occasions some patients may respond to desmopressin. Management should be discussed with the consultant.

2.7. Supportive Care

Rest, bed rest where appropriate. A cold compress will often give good analgesia. Appropriate analgesia starting with Paracetamol and/or Dihydrocodeine but avoiding Aspirin and non-steroidal anti-inflammatory drugs. The COX-2 NSAID’s do not have an anti-platelet effect and may be suitable, or alternatively COX-2 selective NSAID’s.

2.7.1. Physiotherapy

Once an acute joint or muscle bleed has settled then aggressive physiotherapy is necessary to ensure return of full mobility and maintain power around the joint which is essential to it’s stability in protecting the joint from progressive arthritis.

2.7.2. Prophylactic Therapy

- This is used in severe haemophilia A and B.
- The aim is to maintain the factor level >1 iu/dl to prevent spontaneous bleeds and joint destruction.
- It generally commences in childhood, around one to two years of age and is generally continued into adulthood.
- It may also be given in established patients with increasingly troublesome target joints usually for a set period of time, and also in the post-operative period.
- It may be given, for example, as factor concentrate 25-40 iu/kg in haemophilia A (possibly initially once then increasing to 3 times per week) or haemophilia B (generally once or twice weekly).
- Dose requirements should be assessed by trough levels.

Sporadic prophylaxis with either desmopressin or concentrate may also be used in milder patients, before an (often sporting) activity which would normally result in haemorrhage.

2.7.3. Home and community care

Wherever appropriate, the care of patients with haemophilia and their families should be delivered in the home setting which will minimise absence from school and work and help patients to live more effectively with their life long bleeding disorder.

A child with severe haemophilia should be on home treatment by the age of four years at the latest, depending on venous access and family circumstances. The Centre will liaise with patients and their families on home therapy and will monitor the usage of coagulation factor concentrates.

The Centre will undertake a program of education, covering the principles of management of bleeds (and when appropriate prophylaxis), proficiency in venous access, reconstitution and administration of factor concentrate.
Wherever possible, coagulation factor concentrates should be delivered to the patient’s home.

Patients and their families will be educated as to the importance of keeping formal records of all treatments and episodes of bleeding, to enable them to provide the Haemophilia Centre with essential outcome data; the keeping of appropriate records in the home setting will be monitored by the Haemophilia Centre (cf section 3, Monitoring compliance and effectiveness)

2.7.4. Inhibitors
Between 10 and 15% of patients with severe haemophilia A will acquire allo-antibodies to infused factor concentrate resulting in loss of efficacy of treatment. If this is suspected it can be confirmed by specific assay, which may predict response to treatment. Management is initially with increased doses of factor concentrate, bypassing agents such as FEIBA 50-100 IU/kg, or recombinant activated factor VII (r-FVIIa) 270 mcg/kg.

Given the potential considerable cost of treatment if immune tolerance therapy is required, it is necessary to inform the Trust/Commissioners of new patients.

2.8. Acquired Haemophilia
Rarely, acquired haemophilia may be seen, usually in the elderly population, again due to Factor VIII autoantibodies. The bleeding pattern is closer to that of Von Willebrand’s rather than congenital haemophilia. Management is:

Haemostatic:
partly determined by the inhibitor level
Either:
activated prothrombin complex concentrate ie (Baxter Healthcare) FEIBA 50-100/kg max 200u/kg/day),
or
(Novo Nordisk) r-FVIIa eg one to three doses of 90 mcg/kg every 3 hrs or 270mcg/kg stat.

Immunosuppression:
First line: Prednisolone 1mg/kg/d and/or cyclophosphamide eg 50-100mg/d
Second line: High dose immunoglobulin 2g/kg over 2-5 days

If there is no response within 6-8 weeks, second line therapies may be considered. These include Rituximab, and Cyclosporin A, multiple immunosuppressive agents (eg Azathioprine) and modified Malmo or Bonn regimens. and cyclosporin may also be considered for resistant patients

As with inhibitors in congenital patients it is advisable to inform the Trust/Commissioners of new patients.
2.9. Clinic Review
Adults patients are generally reviewed in the Wednesday Haemostasis Clinic to determine patterns of bleeding, revise therapy schedules and assess complications such as arthritis or hepatitis C. Where necessary, multidisciplinary referrals are made.

Dependent on the disease severity review should be offered:

- Mild disease – every 1 year
- Moderate/Severe disease – every 6 months
- Hepatitis C – generally under Consultant Gastroenterologist or every 4-6 months

Arthropathy/physiotherapy
Patients who experience joint bleeds and or have chronic arthropathy need assessment for the range of treatments by physiotherapists, who are responsible for monitoring joint function and improving joints and muscles on a long term basis. Physiotherapy may be required acutely. Joint routine review will be:

- All patients with significant arthropathy - Bi-annual review
- All other patients with severe haemophilia, or minor arthropathy - annual review
- Orthopaedics - review as indicated

Children
The care of severely affected children and young people needs to be influenced by their changing physical and emotional development and as needed requires the regular input of specialist paediatric services.

- Wednesday afternoon (quarterly with Dr Harris)
- Review as above, to include 6monthly joint Clinic with Dr O Tunstall, visiting Paediatric Haematologist

Standard follow-up investigations for patients with bleeding disorders
- Full blood count,
- LFT’s,
- PHLS “save serum”.

Investigations for patients receiving regular factor therapy
As above plus
- Freeze plasma with 6 monthly inhibitor screen – state factor brand received.

Investigations for Patients receiving prophylactic Factor Therapy
As above plus
- Freeze plasma with
- 6 monthly factor "trough" level and inhibitor screen - state factor brand received
- Wherever possible, half-life studies

Hepatitis C (generally the following tests are undertaken by the specialist service)
As per “standard” investigations above plus
- gamma GT,
• alpha feto-protein
• AST,
• INR,
• Consider hepatitis C PCR and US scan.

2.10. Special Circumstances

2.10.1. Surgery
General and dental surgery is always best planned with the Haemostasis Team who will issue a suitable pro-forma (cf Appendix F).

Patients may be managed either by desmopressin, factor infusion, or platelets and where appropriate tranexamic acid as detailed on the pro-forma. When indicated, blood levels are usually taken pre and post-treatment, meanwhile surgery may proceed.

For example in haemophilia and von Willebrand’s disease:

**General surgery**
- target pre-operative factor level:
- Haemophilia A: FVIIIc: 80-100iu/dl
- vWF: FVIIIc: 80-100iu/dl
- vWF:Activity 50 iu/dl

Where appropriate further treatment is determined by levels taken in the post-operative period, thereafter daily.
Regular clinical assessment of haemostasis is crucial
Factor levels should be sustained at least above 50iu/dl, preferably at 100 iu/dl in the immediate post-operative period,
Patients at risk of viral transmission will be identified on the pro-forma.

NB: Desmopressin administration requires the additional prescription of:

Tranexamic acid PO: 25 mg/kg tds for 7 days (adults typically 1g tds) or I.V: 10mg/kg tds
Usually stat injections of concentrate are given, but with major surgery in haemophilia A, a bolus followed by an infusion is given, equating to a dose of 35-50 iu/Kg:

**Bolus** dose iu = patient’s weight x (100 - baseline level) x 0.5

**Infusion** dose rate = 2-3 iu/kg/hr administered via a T tube with 1 litre N Saline over 24 hrs to dilute the factor concentrate to avoid phlebitis.

**Dental Surgery**
The necessity for the intensity of supportive therapy is largely consequent on the requirement for regional anaesthesia. Evidence shows that tranexamic acid mouthwash is highly concentrated in the gums. Hence:
**Dental extraction,**
Where possible this should be undertaken by local infiltration and, or intraligamental anaesthesia,
When a mandibular block is required, overnight observation in hospital may be necessary,
Usually it will be undertaken in the Department of Maxillo-Facial Surgery,
Usually, ie unless a decidual tooth, desmopressin /factor treatment is as outlined for general surgery,
Oral Tranexamic acid (5%) mouth wash is used and possibly tranexamic acid soaked Surgical gauze.

**Restorative therapy**
Often may be undertaken by the patient’s general dentist

Simple local infiltration and, or intraligamental anaesthesia can be managed with:
oral tranexamic 25 mg/kg tds for 7 days (adults typically 1 g tds)

And 5% tranexamic acid mouth wash adults 10 ml, children 2.5-5mls rinse and spit out, 6 hourly for seven days.

2.10.2.  **Hepatitis**
All blood products potentially carry a risk for hepatitis and as a routine patients, over 1 year of age, are vaccinated against hepatitis A and B. Below 1 year of age only hepatitis B eg Engerix B or HBvaxPRO is licensed. Adequate response to hepatitis B should be documented 6 months after the completion of vaccination, if there is an inadequate response advice should be sought. Thereafter a"booster" is given after 5 years which may be sufficient to maintain immunity, with a hepatitis A booster every 10 years.

Of patients treated before 1985, some 90% acquired hepatitis C infection of whom around 20% will develop significant liver dysfunction, including end-stage failure and hepatoma. Specific treatment with Interferon and Ribavirin is administered in consultation with the gastroenterologists.

2.11.  **Data Keeping**
2.11.1.  **New patients**
Those new patients with UKHCDO registerable diseases, should be given an UKHCDO information sheet and asked for their verbal consent to be entered on the central UKHCDO database.

All patients with UKHCDO registerable diseases should be entered on the Departmental excel database (section 2.11.3), at the time of diagnosis, for the following:
- Demographics
- Diagnosis and factor level including severity
- Preferred treatment (1st/2nd/3rd choice)
- Response to treatment (usually Octim)
- HPA Exercise for ’at risk of vCJD for public health purposes
• Date of UKHCDO registration
• Genetics and date tested (where available)
• For haemophilia “female siblings or offspring”

2.11.2. Treatment records
Patients, or their parents, should record of factor usage and bleeding episodes, to enable collation of outcome data for the purposes of on-going care, audit and submissions to Commissioners.

This may be done either with either a Departmental A4 hand written paper record or by Internet based systems (www.haemtrack.nhs.uk).

2.11.3. RCH-T Centre haemostasis patients’ database.
This Excel spreadsheet lists current patients, their diagnosis and treatment of choice. The database is assessable on the Haematology Oncology server at g:/haemophilia/haemostasis patients MMYY.doc (current version). Hard copies should be updated quarterly and circulated to:
• All haematology consultants,
• Dr S Harris, Consultant Paediatrician
• Haemophilia Clinical Nurse Specialist
• Unit managers of Haematology clinic, Lowen, Polkerris, Sennen and Medical Admissions Units
• Unit managers of the Accident and Emergency Departments at RCH and WCH
• Coagulation and Transfusion laboratory seniors

Data collection and collation should be undertaken by the Haemophilia Team (doctors and Haemophilia Clinical Nurse Specialist).

2.12. Carrier detection, genetic counselling and antenatal diagnosis

2.12.1. Haemophilia carriers
• Carriers of haemophilia may have low levels of factor VIII or factor IX and similar clinical problems to patients with mild haemophilia, bleeding following dental or surgical procedures or trauma. These patients may also have particular problems with menorrhagia and bleeding at childbirth
• They may require a range of treatments for the diverse symptoms that they can experience
• Factor VIII or IX assays should be carried out on all carriers of haemophilia. Those who are identified as haemophilia carriers with or without low factor VIII or IX levels should be formally registered. Those with low Factor VIII and IX levels should be reviewed in the same manner as patients with mild haemophilia

2.12.2. Identification of the haemophilia gene and carrier
• genetic counselling should be available before, during and after the process of haemophilia genetic analysis
• a pedigree study should be carried out for each family, to identify obligate carriers, possible carriers and non-carriers and should be constructed using Cyrillic software (g:/families/bleeding/CR number)
• Genetic analysis (intragenic polymorphism and linkage analysis and/or direct gene analysis) should be carried out to establish carriership for female members of the family where there is a patient with haemophilia
• following a diagnosis of carriership, there should be specialised genetic counselling and education, so that carriers can understand the transmission of haemophilia within their own family
• all patients with haemophilia should with written consent have mutation detection carried out

2.12.3. **The haemophilia carrier and pregnancy**
• pregnancy is a potentially serious undertaking in haemophilia as both the mother and infant are at increased risk of bleeding
• formal education about the transmission of haemophilia within the family should be given before starting a pregnancy, if at all possible
• there should be access to an expert fetal medicine unit for discussion of antenatal diagnosis and pre-implantation diagnosis
• a documented care plan for the delivery of any male infant should be established
• there must at all stages of the pregnancy be close collaboration between the obstetric team and the Haemophilia Centre
• post natal confirmation of the diagnosis should be carried out as soon as possible


3. **Monitoring compliance and effectiveness**

<table>
<thead>
<tr>
<th>Element to be monitored</th>
<th>Adherence to the Standard Contract for Haemophilia Service Specification (NHS England 2013) via quarterly reporting to the Specialised Services Quality Dashboard</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lead</td>
<td>Haemostasis MDT</td>
</tr>
<tr>
<td>Tool</td>
<td>Specialised Services Quality Dashboard</td>
</tr>
<tr>
<td>Frequency</td>
<td>Quarterly</td>
</tr>
<tr>
<td>Reporting arrangements</td>
<td>The reports will be discussed at each monthly MDT. Any changes identified in practice will be recommended.</td>
</tr>
<tr>
<td>Acting on recommendations and Lead(s)</td>
<td>Haemostasis MDT</td>
</tr>
<tr>
<td>Change in practice and lessons to be shared</td>
<td>Required changes to practice will be identified and actioned as necessary at each haemostasis MDT A lead member of the team will be identified to take each change forward where appropriate. Lessons will be shared with all the relevant stakeholders</td>
</tr>
</tbody>
</table>

4. **Equality and Diversity**
   1.1. This document complies with the Royal Cornwall Hospitals NHS Trust service Equality and Diversity statement which can be found in the ‘Equality, Diversity & Human Rights Policy’ or the Equality and Diversity website.
4.1. The Initial Equality Impact Assessment Screening Form is at Appendix 2.
Appendix A. Synopsis of the acute management of patients with haemophilia and related conditions

Acute Management:
Assess the site and extent of the bleed
Investigations
Nothing is gained by repeating an APTT, or baseline level, as this will be prolonged as per usual.
For a severe bleed check the full blood count and a citrated sample for “freeze plasma”. Group and screen if indicated, eg haematemesis.
Management is dependent on the nature of the bleed, the deficiency and its severity, documented (if carried) in the patient’s red Bleeding Disorder card”
Refer for confirmation and the appropriate treatment to the RCH-T Haemophilia Centre haemostasis patient database.
Routinely confirm the management and need for further therapy with the on-call Clinical Haematologist
Treat promptly
Determine additional measures eg rest or admission

NB The On call Haematologist should routinely be informed of a patient’s attendance with a bleeding problem and they should never be discharged without treatment unless in consultation with the on-call haematology consultant,

Appropriate treatment - Coagulation Factors should be ordered from Blood Bank on a Transfusion Request Form. Desmopressin should be ordered from pharmacy by it’s brand name OCTIM, out of hours held in the fridges on MAU, ED & Lowen

Haemophilia A - This depends on the phenotypic severity

<table>
<thead>
<tr>
<th>Severity</th>
<th>Baseline factor Level</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>&gt;5</td>
<td>Usually desmopressin (cf. below).</td>
</tr>
<tr>
<td>Moderate</td>
<td>1 – 5</td>
<td>Usually factor concentrate.</td>
</tr>
<tr>
<td>Severe</td>
<td>&lt;1 iu/dl</td>
<td>Always factor concentrate</td>
</tr>
</tbody>
</table>

Haemophilia B
These patients do not respond to desmopressin and require specific FIX concentrate therapy

Von Willebrand’s disease
Most patients have mild type 1 disease and have an excellent response to desmopressin, Desmopressin is best avoided in the treatment of significant head injury.
Type 2 patients have a variable response to desmopressin, whilst this ineffective in the severe type 3 disease, when factor concentrate is required for control of haemorrhage.

Factor concentrate, administration and dosage
recombinant (genetically) manufactured or a plasma derived specific product.
re-constitute with the accompanying diluent as per the manufacturer’s recommendations.
infuse intra-venously, no faster than 10ml per minute and determined by the patient’s comfort.

Clinical Guideline for the management of Primary Haemostasis Disorders
### Diagnosis

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Appropriate choice of product</th>
</tr>
</thead>
<tbody>
<tr>
<td>Haemophilia A (factor VIII deficiency)</td>
<td>Consult excel spreadsheet for individual patients</td>
</tr>
<tr>
<td>Haemophilia B (factor IX deficiency)</td>
<td>Recombinant Factor IX Benefix, or</td>
</tr>
<tr>
<td>Von Willebrand’s Disease</td>
<td>Plasma derived Haemate P.</td>
</tr>
<tr>
<td>other deficiencies, eg FXI,</td>
<td>consult section 4.2.1 of guidance</td>
</tr>
</tbody>
</table>

### Haemophilia A treatment outline

<table>
<thead>
<tr>
<th>Condition/bleed site</th>
<th>Requisite post-treatment factor level for haemostasis</th>
<th>dose of factor VIII iu/kg, when treatment of choice</th>
</tr>
</thead>
<tbody>
<tr>
<td>Minor spontaneous bleeding or joint bleed</td>
<td>30</td>
<td>15</td>
</tr>
<tr>
<td>Severe bleeding, eg muscle or profuse haematuria</td>
<td>40-50</td>
<td>20-25</td>
</tr>
<tr>
<td>Major surgery or <strong>head injury</strong></td>
<td>80-100</td>
<td>35-50</td>
</tr>
</tbody>
</table>

*Significant head injury*, as associated with loss of consciousness, confusion, nausea, deep wound, haematoma or fracture.

**Desmopressin (acetate)** - OCTIM® 15microgrammes/ml synthetic analogue of antidiuretic hormone
boosts factor VIII and von Willebrand factor levels 2 to 4-fold,
give either IV (in 100mls 0.9% Sodium Chloride), or sub-cutaneously once (or twice daily),
activates the fibrinolytic pathways so give tranexamic acid in conjunction,
Best avoided in significant head injury, due to ADH effect,
patients should be water-restricted, to quench their thirst for the following 24 hours.

desmopressin dose = 0.3 micrograms/kg either undiluted SC or IV in 50-100 ml saline over 20 minutes

Tranexamic acid – orally: 25 mg/kg tds for 7 days (adults typically 1 g tds)
I.V: 10mg/kg tds
N.B. Should not be used in patients with haematuria, pregnancy or those with a thrombotic history.

**Additional management:**
In the case of severe trauma or surgery further factor may be administered by bolus or infusion
Joint bleed Consider admission for bed rest or review following day.
Muscle haematoma Admit for bed rest.
**Head injury** Urgent CT. BD therapy, or by continuous infusion - consider levels
Appendix B. Factor concentrate by continuous infusional therapy (See also Clinical guideline for the prescribing and administration of continuous infusion of clotting factor concentrates, available from Document Library)

Although unlicensed, evidence supports the use of infusional therapy, in haemophilia A and B, on the following basis:
Reconstituted factor is stable at room temperature for 24 hours,
Infusional therapy gives steadier haemostatic factor levels, avoiding the peaks and troughs of bolus therapy,
Allows titration of the dose (rate) against the venous factor level,
Generally results in the use of less units of factor.

In the event of any problem or uncertainty, in the first instance contact the Haemophilia CNS, alternatively refer to the nursing staff of Lowen Ward (ext 2050), or the on call Consultant Haematologist.

Procedure
At the instruction of a haematology consultant, who takes responsibility for this care and with reference to the RCH Haematology Department “protocol for factor concentrate in surgery” issued for the patient:

Factor concentrate should be requested, prescribed and administered as follows:

The specified factor concentrate should be requested from the Transfusion Department on the standard form, specifying either the dose or rate* for a defined time period,
The standard infusion rate is 2-3 international units/kg/hr* ,
The specified factor concentrate infusion should be prescribed on the intravenous therapy sheet, specifying the dose, duration and rate ml/hr (see example),
The factor concentrate is reconstituted in the manufacturer supplied diluent
And drawn up into a 50ml syringe for an IVAC P1000 pump with an optimum concentration of 100iu/ml in appropriate solvent
If the infusion is established via a peripheral cannula, administer together 1 litre N Saline over 24 hours (adult), using a multi lumen connector, to dilute the factor concentrate (within the vein) in order to avoid phlebitis.
If the infusion is via a central catheter (eg PICC or Port-a-cath) administer directly into the line.
Example of an initial bolus followed by an infusion with saline flush:

<table>
<thead>
<tr>
<th>Date</th>
<th>Line</th>
<th>Fluid or blood product</th>
<th>Volume</th>
<th>Batch</th>
<th>Duration</th>
<th>Rate</th>
<th>Dr Sign</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 dd/mm/yy</td>
<td>IV</td>
<td>(Brand Name) FACTOR VIII</td>
<td>2000 units</td>
<td>slow bolus</td>
<td>xxx</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2 dd/mm/yy</td>
<td>IV</td>
<td>(Brand Name) FACTOR VIII</td>
<td>2000 units</td>
<td>2ml/hr =200iu/hr</td>
<td>xxx</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3 dd/mm/yy</td>
<td>IV</td>
<td>N Saline flush (via connector)</td>
<td>1000mls</td>
<td>30ml/hr</td>
<td>xxx</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

NB
Following surgery or a significant bleed or procedure the target level is 100 iu/dl
Unless instructed, the infusion must be continuous, with the syringe replenished,
The ongoing infusion rate is dependant on the steady state factor level, a citrate sample (sky blue bottle) taken by venepuncture from the opposite limb, usually undertaken post-operatively/procedure and thereafter every 24hrs,
Flush through rather than discard infusion lines (a dead space of 1ml contains a significant number of units)
Appendix C: Physiotherapy review

<table>
<thead>
<tr>
<th></th>
<th>Acute*</th>
<th>Routine</th>
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<tbody>
<tr>
<td><strong>Adults</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Severe haemophilia/vWD, or those with significant arthropathy</td>
<td>As required</td>
<td>Bi-annual</td>
</tr>
<tr>
<td><strong>Adults</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moderate/mild haemophilia</td>
<td>As required</td>
<td>if there is a target joint or arthropathy - Annually otherwise - as required,</td>
</tr>
<tr>
<td><strong>Children</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Severe haemophilia</td>
<td>As required</td>
<td>Bi-annual</td>
</tr>
<tr>
<td><strong>Children</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moderate/mild haemophilia</td>
<td>As required</td>
<td>if regular bleeding - annually otherwise - as required</td>
</tr>
</tbody>
</table>

*In-patient referral should be seen within 24-48 hrs.

Pre-pregnancy
All patients with haemophilia A & B to have genealogy completed
Offer counselling and phenotypic/genetic diagnosis to potential carriers
Offer follow up for carriers and partners considering pregnancies

Antenatal management

Confirmation of suspected family history and counselling
At booking, refer known or possible carriers to Haematology Dept for:
Confirmation of family bleeding disorder
Counseling regarding the diagnosis and its implications
antenatal diagnosis

Establish the risk for an affected fetus
As indicated arrange phenotypic/genetic diagnosis by cordocentesis or CVS sexing (20 weeks) by USS of known or potential carriers

Determination of maternal risk of bleeding:
Factor levels at 32-36 weeks of gestation
Peri-partum management plan, covering possible use of desmopressin, or as necessary (recombinant) factor concentrate, if difficult labour, borderline levels, or unexpected bleeding.

Plan for delivery:
Delivery of a possible affected case should be at RCH, unless otherwise agreed
Delivery route should be on the basis of obstetric indications
But avoid ventouse, instrumental delivery and fetal scalp monitoring

Management of neonate:
Obtain urgent 5ml cord (sky blue citrated) or 1ml venous blood (blue paediatric bottle) sample for coagulation screen/ specific factor analysis
Inform Consultant Haematologist of birth
Avoid IM injections
Give oral vitamin K

In an affected neonate:
If traumatic delivery, consider specific factor concentrate
Consider routine USS to exclude ICH at day 3.
Consider early hepatitis B vaccination (by SC route)
Blood tests by heel prick sampling should be taken with great care (pressure for 3 minutes)
Arrange follow up in joint Paediatric Haemostasis Clinic
Management plan: peri-partum care for carriers of haemophilia

Patient details/sticker:

Name: 
Hospital no: 
Address: 

This woman is a carrier of haemophilia. Her latest factor VIII level is taken on ……………. Although there is a risk of bleeding if her factor VIII level is low, prophylactic treatment is not recommended if the level is >50iu/dl (normal non-pregnant range 50-150). Anaesthetic advice (Dr.) is that she is suitable for regional analgesia as long as her factor VIII level is normal (>50iu/dl).

There is a 50% risk of her baby having haemophilia. The aim of this plan is to achieve an atraumatic delivery and avoid risk of fetal/neonatal haemorrhage.

Additional points:
- Coagulation and factor VIII levels will have been checked in third trimester and will usually have risen 3-4 fold

Care of the mother in labour:
- Delivery at RCH, (please inform on call Consultant Obstetrician)
- Delivery route based on obstetric indications
- Avoid prolonged 1st or 2nd stage
- Avoid instrumental delivery
- Avoid fetal scalp electrode
- Avoid scalp sampling, but if necessary ensure pressure applied at site for at least 5 mins
- Repair any perineal trauma immediately
- Early and aggressive treatment of any post-partum haemorrhage
- In the case of difficult labour, borderline factor VIII levels, unexpected bleeding, consider the use of desmopressin (OCTIM® 15microgrammes/ml in drug cupboard on Delivery Suite; 0.3microgrammes/kg in 50ml N/saline over 20 minutes) or specific factor concentrate

Care of the neonate:
- Citrated sample of cord blood (sky blue top) URGENTLY to haematology for coagulation screen/factor VIII analysis
- Inform Consultant Haematologist of delivery
- NOT for intramuscular injections
- Oral vitamin K
- If delivery traumatic, consider specific factor concentrate
- Consider routine ultrasound day 3 to exclude ICH
- Consider early hepatitis B vaccination (sub-cutaneous route)

Date:
Signed:
Appendix E: Vaccinations and injections In Children with bleeding disorders

Children with Haemophilia, Von Willebrand's disease and other bleeding disorders can get painful muscle bleeds (haematomas) if given intramuscular injections. Alternative routes are preferred:
- Subcutaneous injection for most vaccinations
- Oral vitamin K following birth
- Intradermal injection for BCG

Vitamin K

Vitamin K deficiency may occur in the first few months of life and can lead to serious bleeding (Haemorrhagic disease of the newborn). Babies in the UK are offered Vitamin K to prevent this, usually by intramuscular injection. If given orally several doses are needed because it is not so well absorbed into the body compared to intramuscular injection. Doses are given:
- Following birth
- Second dose at 4-7 days
- Third dose at 1 month and each subsequent month if exclusively breast-fed. (Third and subsequent doses not required if on formula milk or weaned)

Vaccinations

Routine childhood vaccinations should be given by the subcutaneous route, except BCG, which is given intradermally. MMR vaccination should be given deep subcutaneous injection. These vaccinations can be done at the usual recommended ages and can be done in the GP surgery or school.

All children who receive or are likely to receive blood products should also receive vaccinations against Hepatitis A and B viruses. All blood donors and blood products are screened for known viruses so the risk of these illnesses being transmitted through blood is very small.

Hepatitis B

This can be given at any age. There are several forms e.g.
- Engerix B 0.5mls (10 microgms) at birth to 1 month with second and third doses given 1 month and 2 months after the first injection, and a forth dose at 12 months. For children over 12 years the dose is 1ml. A booster may be required 5 years after the primary course. HBvaxPRO is another Hepatitis B vaccination.
- Other dosage regimes may be recommended in certain circumstances.

Hepatitis A

This is licensed to give in children over 1 year (although may be given before the age of one if necessary). Several forms are available e.g. Havrix Junior Monodose 0.5mls at 1 year of age and booster dose 6-12 months after.
- A combined Hepatitis A and B vaccination is available for children over 1 year - Twinrix, (0.5mls for children aged 1-15yrs) which involves a second and third injection 1 month and 6 months after the first.

Hepatitis A and B vaccinations are inactivated so there is no time interval required between these vaccinations and the routine childhood vaccinations.
Appendix F: List of additional documents, pro-formas and templates:

The following guidance and pro-formas are available on the Haematology Oncology server (under k:/protocols and specific diseases/haemostasis/)

Management of primary disorders of haemostasis
Protocol for factor concentrate management in surgery
Protocol for use of desmopressin management in surgery
Desmopressin and Tranexamic acid patient information
Clinical guideline for the prescribing and administration of continuous infusion of clotting factor concentrates

Template letters are also available for

Clinic call-up
Holiday letter
Screening siblings

As needed these are available through the secretaries
Appendix G: RCHT Pathway for Out of Hours Assessment Treatment and Follow Up of Patients With Primary Haemostatic

Patient Requires Out of Hours Service

- Patient has follow up appt in location arranged by Haemophilia Centre
- Patient phones for telephone advice
- Patient presents directly to ED
- Patient calls an ambulance
- Patient presents directly to HO Clinic

- Given telephone advice only
- Advised to go to Truro ED, H/O, Paed Ward or call an ambulance. If on home treatment should be advised to self administer treatment prior to
- Advice given & documented. Haemophilia Centre contacted next working day to arrange appropriate follow up
- Patient triaged within 30 mins
- Patient assessed, Consultant Haematologist on call contacted via switchboard. Plan agreed, treatment given, scans, xrays
- Patient discharged home
- Patient admitted
- Next working day Haemophilia Centre informed of presentation
- Follow up arranged by Haemophilia Team

Regular Audit of Service

Haemophilia Director: Dr M D Creagh (ext 2524 or netpage via switchboard)
Haemophilia CNS: Sarah Johns (ext 3239 or 07817134678)
Haematologist On Call: netpage via switchboard
Lowen Ward: 01872 252050
Appendix H: Availability of Coagulation Factor Concentrates

Whilst preferable not to change product for a patient on regular treatment*, where treatment is necessary (usually for a holidaymaker) it is acceptable to substitute the patient's usual product for one held in stock at RCHT.

*Many visitors with a severe bleeding phenotype will carry their own emergency (initial) supply of factor

<table>
<thead>
<tr>
<th>Type of Product</th>
<th>Indication for use</th>
<th>Products held in stock at RCHT</th>
<th>Brand names of other products</th>
</tr>
</thead>
<tbody>
<tr>
<td>Recombinant Factor VIII</td>
<td>Moderate &amp; Severe Haemophilia A</td>
<td>Refacto AF (Wyeth)</td>
<td>Kogenate (Bayer)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Advate (Baxter)</td>
<td>Helixate (CSL Behring)</td>
</tr>
<tr>
<td>Plasma Derived Factor VIII (intermediate purity)</td>
<td>Moderate &amp; Severe Haemophilia A</td>
<td>Haemate P (CSL Behring)</td>
<td>8Y (BPL)</td>
</tr>
<tr>
<td></td>
<td>Type 2 &amp; 3 von Willebrand Disease which is unresponsive to desmopressin or where desmopressin is contraindicated</td>
<td></td>
<td>Alphanate (Grifols)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Wilate (Ocatpharma)</td>
</tr>
<tr>
<td>Recombinant Factor IX</td>
<td>All types of Haemophilia B</td>
<td>Benefix (Wyeth)</td>
<td>NA</td>
</tr>
<tr>
<td>Plasma Derived Factor IX</td>
<td>All types of Haemophilia B</td>
<td>Not held in stock</td>
<td>Mononine (CSL Behring)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Replinine VF (BPL)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Alphanine (Grifols)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Haemonine (Biotest UK)</td>
</tr>
<tr>
<td>Bypassing Agents</td>
<td>All inhibitor patients</td>
<td>FEIBA (pd) (Baxter)</td>
<td>NA</td>
</tr>
<tr>
<td></td>
<td></td>
<td>NovoSeven (recombinant) (NovoNordisk)</td>
<td></td>
</tr>
<tr>
<td>Activated Prothrombin Complex Concentrates (APCC’s)</td>
<td></td>
<td>Beriplex (CSL Behring)</td>
<td>Octaplex (Octapharma)</td>
</tr>
</tbody>
</table>
Appendix I: References and links

These guidelines are drawn together with reference to published guidance from the UK Haemophilia Centre Doctors’ Organisation (UKHCDO).
http://www.ukhcdo.org/UKHCDOguidelines.htm

Emergency and out of hours care for patients with bleeding disorders – Standards of care for assessment and treatment.(2010) UKHCDO website

Guideline on the selection and use of therapeutic products to treat haemophilia and other hereditary bleeding disorders. A United Kingdom Haemophilia Center Doctors’ Organisation (UKHCDO) guideline approved by the British Committee for Standards in Haematology Haemophilia, 14, 671-684

The diagnosis and management of von Willebrands Disease: a UKHCDO guideline approved by the BCSH (BJH 2014) doi10.111/bjh13064 http://www.blackwellpublishing.com

Guideline for the Diagnosis and Management of the Rare Coagulation Disorders
Haemophilia, Volume 19, Issue 3, pages e191–e192, May 2013


Diagnosis and management of acquired coagulation inhibitors: a guideline from UKHCDO Br J Haematol, 162:758-773, 2013


Haemorrhagic disorders in pregnancy – British Committee for Standards in Haematlogy 1994

Neonatal haemostasis and thrombosis - British Committee for Standards in Haematlogy 2002

The Haemophilia Society offers support and patient information at http://www.haemophilia.org.uk
Appendix J: Members of the Haemostasis Team

Consultant and Haemophilia Director: Dr M D Creagh Ext 2503 or netpage
Consultant Haematologists: Dr J Blundell Ext 2507 or netpage
Dr A R Kruger,
Dr R S Noble,
Dr BT Pottinger,
Dr E Parkins
Dr A Forbes

Haematology SPR netpage
Haemophilia Clinical Nurse Specialist: Ms S Johns 07880 502491
Consultant Paediatrician Dr S Harris Ext 2728 or netpage
Consultant Paediatric Haematologist Dr O Tunstall Bristol Hospital for Sick Children tel 0117 3428752

Secretary Ms Kerry Morris Ext 2524

BMS 3 Transfusion Department Mr S Bassey Ext 2500
Clinical Scientist Coagulation section Mr P Carson Ext 2052
Mrs C Errington Ext 2052

Liaison Physiotherapist: Sophie Hendriksz/Eva Dingwall

And

Consultant Hepatologist Dr H Hussaini
Hepatitis Clinical Nurse Specialist Mrs L Farrington
Consultant Rheumatologist Dr D Hutchinson
Orthopaedic Consultants:
Mr D Bracey
Mr M Regan
Mr D Fern

Maxillo-Facial Surgeons: Mr C Lansley
Mr S Addock
Mr M Retallick (Associate Specialist)

Adult patient Specialist on call advice:
Oxford Comprehensive Care Centre
Churchill Hospital Tel 01865 741166
# Appendix 1. Governance Information

<table>
<thead>
<tr>
<th>Document Title</th>
<th>Clinical Guideline for the management of Primary Haemostasis Disorders (Haemophilia &amp; Related Conditions)</th>
</tr>
</thead>
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<td>May 2015</td>
</tr>
<tr>
<td>Date Valid From:</td>
<td>May 2015</td>
</tr>
<tr>
<td>Date Valid To:</td>
<td>May 2018</td>
</tr>
<tr>
<td>Directorate / Department responsible (author/owner):</td>
<td>Dr MD Creagh, Consultant Haematologist &amp; Haemophilia Director</td>
</tr>
<tr>
<td>Contact details:</td>
<td>01872 252524</td>
</tr>
<tr>
<td>Brief summary of contents</td>
<td>Guidance for the care and management of patients with Primary Haemostatic Disorders at RCHT</td>
</tr>
<tr>
<td>Suggested Keywords:</td>
<td>Factor 8, Factor VIII, Factor 9, Factor IX Haemostasis, Haemophilia, von willebrand disease, bleeding</td>
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<td>Target Audience</td>
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<td>Medical Director</td>
</tr>
<tr>
<td>Date revised:</td>
<td>April 2015</td>
</tr>
<tr>
<td>This document replaces (exact title of previous version):</td>
<td>Guidance for the Management of Primary Haemostatic disorders (Haemophilia and Related Conditions) V1.0</td>
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<td>Haemostasis MDT; CSSC Governance DMB</td>
</tr>
<tr>
<td>Divisional Manager confirming approval processes</td>
<td>Sally Rowe, Divisional Director CSSC</td>
</tr>
<tr>
<td>Name and Post Title of additional signatories</td>
<td>Janet Gardner, Governance Lead CSSC</td>
</tr>
<tr>
<td>Signature of Executive Director giving approval</td>
<td>{Original Copy Signed}</td>
</tr>
<tr>
<td>Publication Location (refer to Policy on Policies – Approvals and Ratification):</td>
<td>Internet &amp; Intranet</td>
</tr>
<tr>
<td>Document Library Folder/Sub Folder</td>
<td>Clinical/Haematology</td>
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Links to key external standards

<table>
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<th>Links to key external standards</th>
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Related Documents:

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<th>Related Documents:</th>
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<tr>
<td>See reference list at Appendix I</td>
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Training Need Identified?

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Version Control Table

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<th>Summary of Changes</th>
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<tbody>
<tr>
<td>Jan 2012</td>
<td>V1.0</td>
<td>Initial Issue, in new Trust Format</td>
<td>Dr MD Creagh, Haemophilia Director</td>
</tr>
<tr>
<td>April 2015</td>
<td>V2.0</td>
<td>Updated standards in line with new national contracts.</td>
<td>Dr MD Creagh, Haemophilia Director</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Updated guidance in line with recent publications.</td>
<td>Sarah Johns, Haemostasis CNS</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Rewritten to fit Trust Clinical Guideline format</td>
<td></td>
</tr>
</tbody>
</table>

All or part of this document can be released under the Freedom of Information Act 2000

This document is to be retained for 10 years from the date of expiry.

This document is only valid on the day of printing

Controlled Document

This document has been created following the Royal Cornwall Hospitals NHS Trust Policy on Document Production. It should not be altered in any way without the express permission of the author or their Line Manager.

Clinical Guideline for the management of Primary Haemostasis Disorders

Page 31 of 33
## Appendix 2. Initial Equality Impact Assessment Form

<table>
<thead>
<tr>
<th>Name the strategy / policy / proposal / service function to be assessed (hereafter referred to as <em>policy</em>) (Provide brief description): Clinical Guideline for the management of Primary Haemostasis Disorders (Haemophilia &amp; Related Conditions)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Directorate and service area: CSSC, Haematology</td>
</tr>
<tr>
<td>Name of individual completing assessment: Sarah Johns</td>
</tr>
<tr>
<td>1. Policy Aim* Who is the strategy / policy / proposal / service function aimed at?</td>
</tr>
<tr>
<td>2. Policy Objectives*</td>
</tr>
<tr>
<td>3. Policy – intended Outcomes*</td>
</tr>
<tr>
<td>4. How will you measure the outcome?</td>
</tr>
<tr>
<td>5. Who is intended to benefit from the policy?</td>
</tr>
<tr>
<td>6a) Is consultation required with the workforce, equality groups, local interest groups etc. around this policy?</td>
</tr>
<tr>
<td>b) If yes, have these *groups been consulted?</td>
</tr>
<tr>
<td>C). Please list any groups who have been consulted about this procedure.</td>
</tr>
</tbody>
</table>

### 7. The Impact

Please complete the following table.

<table>
<thead>
<tr>
<th>Equality Strands:</th>
<th>Yes</th>
<th>No</th>
<th>Rationale for Assessment / Existing Evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>X</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
### Clinical Guideline for the management of Primary Haemostasis Disorders

**Gender** (male, female, transgender / gender reassignment)
- X

**Race / Ethnic communities / groups**
- X

**Disability** -
- learning disability, physical disability, sensory impairment and mental health problems
- X

**Religion / other beliefs**
- X

**Marriage and civil partnership**
- X

**Pregnancy and maternity**
- X

**Sexual Orientation, Bisexual, Gay, heterosexual, Lesbian**
- X

You will need to continue to a full Equality Impact Assessment if the following have been highlighted:
- You have ticked “Yes” in any column above and
- No consultation or evidence of there being consultation - this excludes any policies which have been identified as not requiring consultation. or
- Major service redesign or development

8. Please indicate if a full equality analysis is recommended. | No

9. If you are not recommending a Full Impact assessment please explain why.

Does not meet criteria as above.

<table>
<thead>
<tr>
<th>Signature of policy developer / lead manager / director</th>
<th>Date of completion and submission</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Names and signatures of members carrying out the Screening Assessment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Dr M D Creagh</td>
</tr>
<tr>
<td>2. Sarah Johns</td>
</tr>
</tbody>
</table>

Keep one copy and send a copy to the Human Rights, Equality and Inclusion Lead, c/o Royal Cornwall Hospitals NHS Trust, Human Resources Department, Knowledge Spa, Truro, Cornwall, TR1 3HD

A summary of the results will be published on the Trust’s web site.

Signed _Dr M D Creagh_

Date 30.04.15__