Hereditary Bleeding Disorders - Full Clinical Guideline - Derby Sites Only

Reference no.: CG-T/2025/175

1. Introduction

Royal Derby Hospital is a Haemophilia treatment centre. This means the haematology department diagnoses and treats people (adults and children) with hereditary bleeding disorders. This includes Haemophilia A and B, von Willebrands disease and hereditary platelet disorders. Also rare bleeding disorders such as factor XI, Factor X and Factor VII deficiency, and fibrinogen deficiency.

When persons with Haemophilia, von Willebrands disease or other hereditary bleeding disorders present to UHDB NHS Trust the haematology consultant/SpR must be contacted immediately:

Monday – Friday, 9 to 5: Haemostasis CNS 07469405987 or telephone haematology secretaries on ext. 87973

Other times and Bank Holidays: Contact the hospital switchboard and ask for the Haematology Consultant or Registrar on-call.

Diagnostic or therapeutic interventions, Surgery, Trauma, Spontaneous Bleeds.

Note: even minor interventions e.g. Arterial Blood Gas, lumbar puncture, arterial line or central venous catheter placement, solid organ biopsy (kidney or liver), chest drain placement etc… can lead to bleeding. This is not an exhaustive list.

A management plan for managing the bleeding disorder must be completed by the Haemostasis consultant (Dr A Mckernan or Dr M Ruparellia (or other consultant haematologist/registrar) before any planned procedure. For emergency procedures the on-call Haematology Consultant must decide on the management plan. See Appendix ‘Operation plan template for patients with hereditary bleeding disorders’

2. Factor VIII – clotting factor 8; CCC – Comprehensive Care centre; VWD – von Willbrands disease; vCJD – variant creutzveldt Jacob disease; iu/dl = %

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Introduction

Derby hospitals is a Haemophilia treatment centre. This means the haematology department diagnoses and treats people (adults and children) with hereditary bleeding disorders. This includes Haemophilia A and B, von Willebrands disease and hereditary platelet disorders. Also rare bleeding disorders such as factor 11 deficiency , factor 10 deficiency , factor 7 deficiency , and fibrinogen deficiency.

With the exception of the rare bleeding disorders (for which see page 16), the commoner hereditary bleeding disorders (Haemophilia and von Willebrands) may be divided into two main groups:

Mild bleeding disorders responsive to desmopressin – this includes the vast majority of patients with von Willebrands disease, and some patients with mild haemophilia and platelet disorders. **THESE PATIENTS DO NOT USUALLY REQUIRE CLOTTING FACTOR CONCENTRATE.**

Severe and moderate bleeding disorders – these patients require treatment (often frequently) with clotting factor concentrate (CFC). The patients who require CFC are listed on the Haemophilia Register.

Haemophilia Register

A copy of the haemophilia register is kept on the Haematology shared drive. The register contains the patient details, diagnosis, factor level and the clotting factor concentrate needed to treat each patient.

Visiting patients.

Patients who are passing through our area may present to us and will not be on our haemophilia register. As well as taking a thorough history from the patient about their bleeding disorder and treatment always contact their own Haemophilia Centre to confirm the treatment and management. Be wary of mild bleeders who are infrequently treated. They may not be aware of what treatment they have had. When asked directly they may say they have had clotting factor concentrate when in fact it was Desmopressin (see Previously Untreated Patients below)

Treatment areas.

Patients presenting Monday – Friday 9 – 5 are seen in the following places:
Adults; Combined Day Unit/Specialist outpatients
Children; childrens emergency department.

Other times:
Adults: A and E
Children: Childrens A and E, Puffin ward.

Comprehensive Care Centre (CCC)

QMC, Nottingham is our CCC. Other centres worth contacting for advice are or Sheffield Royal Hallamshire, Birmingham QE. , .

Clotting factor concentrate (CFC)

Clotting factor concentrate (CFC) is stocked in Pharmacy **(however**, **for emergency use a small amount is kept in 2 places around the Trust see page 5).**

The products we stock are:
Haemophilia A
ADVATE (recombinant FVIII)
Haemophilia B
BENEFIX (recombinant FIX)

FXIII deficiency
FIBROGAMMIN (plasma derived FXIII)

FX deficiency
OCTAPLEX (plasma derived concentrate containing FX, FII, FVII, and FIX, also used for reversing warfarin)

Acquired FVIII inhibitor
FEIBA (plasma derived Factor VIII Inhibitor Bypassing Activity – similar to Octaplex but with the factors deliberately activated).
Obizur (recombinant porcine factor VIII)
Novoseven. Recombinant VIIa.

Type 2 von Willebrands disease (the vast majority of von Willebrands disease patients have type 1 disease, respond to desmopressin and should not usually need clotting factor concentrate)
Wilate (plasma derived FVIII and VW concentrate)
Veyvondi (recombinant VW concentrate - FVIII levels will take up to 12 hours to normalize so for an acute bleed one dose of FVIII concentrate needs to be given).

Pharmacy keep the main stock but for emergency use a small amount is kept in 2 places around the Trust:

1. **Blood issue fridge room on the 5th Floor**, next door to blood bank: combination for fridge is 2244.
Refacto (recombinant factor 8 for haemophilia A): 5000 units
Advate, (recombinant factor 8 for haemophilia A): 5000 units
Benefix (recombinant factor 9 for haemophilia B): 10,000 units
Wilate (von Willebrand and Factor 8, plasma derived, for type 2 VWD): 5000 units

2. **Childrens emergency dept** - in a fridge in the room at the back of children's' resus:
Refacto and advate - 5000 units each
Wilate 2000 units.

If clotting factor is not available in Derby, contact Nottingham Haemophilia Centre or the relevant company.

Principles of treatment.

Local Measures:
Where possible use local haemostatic measures to control bleeding. e.g pack and suture tooth socket, pack/cauterize nose, topical tranexamic acid for mouth bleeding.

Antifibrinolytics:
Unless contraindicated, give Tranexamic acid orally or IV (starting 2 hours before any planned procedure). For adults the dose is 1 gram QDS, for children consult paediatric BNF.

Increase the clotting factor level (if necessary):
Treatment depends on which clotting factor is deficient, how severe the deficiency is and what level is required. If clotting factor is necessary: WHEREVER POSSIBLE RECOMBINANT PRODUCTS SHOULD BE USED IN PREFERENCE TO PLASMA DERIVED PRODUCTS. See below.

Diagnostic or therapeutic interventions, Surgery, Trauma, Spontaneous Bleeds.

Note: even minor interventions e.g. Arterial Blood Gas, lumbar puncture, arterial line or central venous catheter placement, solid organ biopsy (kidney or liver), chest drain placement etc… can lead to bleeding. This is not an exhaustive list.

A management plan for managing the bleeding disorder must be completed by the Haemostasis consultant/Senior Clinical Fellow (or other consultant haematologist/registrar) before any planned procedure. For emergency procedures the on-call Haematology Consultant must decide on the management plan. See Appendix 'Operation plan template for patients with hereditary bleeding disorders'

Mild bleeding disorders responsive to desmopressin.

e.g. Most Von Willebrands disease (VWD), mild haemophilia A with FVIII > 20%, carriers of Haemophilia A (1/3 have slightly low FVIII levels), some platelet disorders. **Haemophilia B does not respond to desmopressin.**

Desmopressin can be given subcutaneously or intravenously. Peak VW and FVIII levels occur at 1 hour post injection. Response lasts 12 – 24 hours. The effect may wear off sooner than 12 hours. A repeat dose may be given but may cause fluid retention and hyponatraemia.

Mild haemophilia A.

The term 'Mild' haemophilia A covers FVIII levels from 6% - 40%.

Desmopressin increases the FVIII level 2 - 3 fold. For factor levels towards the lower end of that range i.e. < 15%, clotting factor will be necessary. For 15% and above desmopressin should be considered first. Depending on the factor VIII level and the severity of the bleed/operation desmopressin may suffice.

e.g. Baseline of 6%: Desmopressin will increase the FVIII to probably 20%. These people will probably need clotting factor.

Baseline 15%. Anyone with 15% or more FVIII should be considered for desmopressin. Desmopressin will increase the factor VIII to approx 50%, enough for mild bleeds or minor procedures. For major bleeds/procedures clotting factor may be necessary.

Baseline 30-49%. Desmopressin will increase FVIII into the normal range and in most to 100%. Check if the patient has had a documented response in the past. Bear in mind that trauma/stress will increase the baseline. With major procedures/trauma stress my maintain the FVIII in the normal range without out further treatment.

If a second dose of desmopressin is necessary fluid balance and sodium need close monitoring. In mild haemophilia the response to repeat doses of desmopressin gets less - tachyphylaxis- if this happens it may then be necessary to give clotting factor).

Always take FVIII levels pre and post treatment (post is 1 hour after desmopressin, 20 mins after concentrate). We can do FVIII levels as an emergency when necessary. If not necessary samples can be frozen and done the next day.

Von Willebrands Disease.
Most people with VWD will respond to Desmopressin. Desmopressin is given either subcutaneously or IV. FVIII, Ricof (functional VW factor) and closure time are done before and after administration of desmopressin to assess the response if a response has not been documented before or for major bleeding/surgery.

Platelet function disorders.
Some of these respond to desmopressin. If not it may be necessary to use platelet transfusion. Avoid platelet transfusion if possible. This must be discussed with a consultant haematologist. If platelets are used they should be HLA matched.

GUIDELINES FOR THE **SUBCUTANEOUS** ADMINISTRATION OF DESMOPRESSIN ('OCTIM injection') FOR THE TREATMENT OF VON WILLEBRANDS DISEASE, MILD HAEMOPHILIA and PLATELET FUNCTION DISORDERS. **NB children who need desmopressin should receive it IV, see below.**

Desmopressin

- This is a synthetic analogue of Vasopressin which is given by subcutaneous injection.
- It increases the patients' own Factor VIII and von Willebrand factor levels by stimulating the release of these factors from the lining of blood vessels.
- Blood tests are taken before and 60 minutes after subcutaneous injection to monitor the response.
- When Desmopressin is being given for the first time blood tests are also taken six hours after the injection. This may also be done for major surgery.
- Desmopressin may be given as a 'one-off' e.g. as a therapeutic trial to assess the response, or to cover a minor surgical procedure.
- It may also be given daily for 2 – 3 days to cover major surgery – in this case the frequency of administration is guided by monitoring of blood tests.
- Unless contraindicated Tranexamic acid 1 gram QDS orally (or 10mg/kg IV) should be given for 5 days starting at the same time as the Desmopressin.

Side effects.
Hypotension and facial flushing are not uncommon.
Fluid retention and hyponatraemia can occur with repeated doses.

Cautions:
Use with caution in children under 2 years.
Avoid in patients with heart failure, other conditions being treated with diuretics, and those with known atherosclerosis.

Administration of Desmopressin:

Preparation: OCTIM injection, 15 microgram per ml (1 ml PER vial).

Dose: 0.3 micrograms per Kg (capped at 25 micrograms) given by subcutaneous injection.

Monitoring: Check pulse and BP for 1 hour after administration..
In-patients – start fluid balance chart

Blood tests: See below

Fluid restriction: The patient should be advised to restrict their fluid intake to 1 L for 24 hours after the infusion. 'Drink when thirsty' i.e, partial fluid restriction.

If IV fluids are given perioperatively, fliud balance, body weight and serum sodium must be closely monitored.

If a repeat dose of Desmopressin is given fluid balance, body weight and serum sodium must be closely monitored.

Unless contraindicated Tranexamic acid 1 gram QDS orally (or 10mg/kg IV) should be given for 3 - 5 days preferably starting the day before.

Blood tests.

Before the injection:
Take one purple, one red and two blue top bottles for:
FBC, FVIII, VW function, VW Ag, Closure Time, U and E.

60 minutes after the injection (timing is important):
Take one purple and two blue top bottles for:
FBC, FVIII, Ricof, Closure Time.

Patients receiving a test dose of desmopressin or having major surgery:
6 hours after the injection:
Take 2 blue top bottles for:
FVIII, Ricof and Closure Time.

Patients receiving repeated doses of Desmopressin:
U and E (bottle - one red top) must be done at least daily.

Patients having one dose of desmopressin for a minor procedure who have a documented response to desmopressin:
Blood tests not necessary.

Haemophilia.
Patients with haemophilia do not need to have VW levels or Closure Time tests. All other advice applies.

SEVERE AND MODERATE BLEEDING DISORDERS.

Diagnostic or therapeutic interventions, Surgery, Trauma, Spontaneous Bleeds.

Note: even minor interventions e.g. Arterial Blood Gas, lumbar puncture, arterial line or central venous catheter placement, solid organ biopsy (kidney or liver), chest drain placement etc… can lead to bleeding. This is not an exhaustive list.

A management plan for managing the bleeding disorder must be completed by the Haemostasis consultant (or other consultant haematologist/registrar) before any planned procedure. For emergency procedures the on-call Haematology Consultant must decide on the management plan. See Appendix 'Operation plan template for patients with hereditary bleeding disorders'

Severe and moderate Haemophilia A (FVIII < 1% and 1 – 5% respectively). **Mild haemophilia A with FVIII < 20%.**
For Haemophilia B see below (dose formula and FIX half life is different).

CFC necessary. The normal range for FVIII is 50 – 100 iu/dl. The half life of factor VIII is 12 hours.

Calculating the dose of clotting factor concentrate.
The dose of FVIII needed to achieve the desired increase in FVIII level is calculated by the following formula

$$\frac{\text{(Desired rise in FVIII – baseline FVIII) x wt (kg)}}{2} = \text{Units of FVIII}$$

So, for a 70 kg pt with severe haemophilia A (baseline FVIII < 1%), give 3,500 units FVIII to increase level to 100% (round up to whole vial – don't waste part of a vial)

The desired increase depends on the severity of bleed.

For major bleeds or surgery blood samples (blue top bottles) should be taken immediately pre and 20 minutes post the first injection of CFC (and if necessary repeated daily). In some circumstances it may not be necessary to do the levels immediately but the samples can be frozen to be done the following day. Always contact the haematology lab to let them know the samples are coming and whether the results are needed immediately. Samples must be sent or taken to the lab urgently in either case. (See surgery guidelines below).

Treatment of bleeds.
Major bleeds e.g head injury, muscle bleed, large joint bleed (the joint is swollen)– increase FVIII to 100% using the above formula. Depending on the severity of the bleed it may be necessary to keep the FVIII level > 50% for a few days (see below). If not repeat a daily dose of 50 – 100% for 2 or 3 days.

Psoas muscle bleed. Symptoms are of groin pain and difficulty walking, signs are inability to fully extend the hip, numbness over the anterior thigh, possible loss of knee reflex. Elevate FVIII to 100% and maintain > 50% for up to 1 week.

Joint bleed: Elevate FVIII to 100%, repeat 50 – 100% next day, 50% the following day.

For patients not on prophylaxis, a period of prophylaxis may be necessary after any of the above.

Mucosal bleeding (nose or mouth bleeding) give one dose of FVIII of 50%, plus tranexamic acid (1 gram QDS for 5 days). Topical tranexamic acid may be used: soak gauze in IV preparation, mouthwash)

Painless haematuria – increase fluid intake, avoid CFC if possible (risk of clot colic).

In some circumstances it may be necessary to keep the FVIII level in the normal range for several days e.g. psoas bleed or other major muscle bleed:

The half-life of FVIII is 12 hours (in steady state; reduced after major surgery, infection), so BD dosing is necessary to maintain FVIII levels > 50% (i.e. in normal range).

After the initial 100% dose is given, a 50% dose is given 12 hours later and then at 12 hourly intervals for as long as necessary. In this situation FVIII levels are done immediately before and 20 mins after the morning dose. The results will help fine tune the dose of FVIII.

Diagnostic or therapeutic interventions:
Even minor interventions e.g. Arterial Blood Gas, lumbar puncture, arterial line or central venous catheter placement, solid organ biopsy (kidney or liver), chest drain placement etc… can lead to bleeding. This is not an exhaustive list.

For all of the above, and any similar procedures, normalize the clotting factor level and maintain in the normal range for 36 – 48 hours, then review.

Haemophilia B (factor IX deficiency). All the above applies except that desmopressin does not work, and the formula for dosing is:

Desired rise in FIX x body wt in Kg = Units of FIX.
To achieve a 100 % rise in factor IX for a 70 kg person needs a dose of 7000 units of FIX.

The half-life of FIX is 18 – 30 hours so repeated dosing is done approximately once daily but may be less (e.g. repeat dose needed after 18 hours)

Administration of CFC
CFC comes as vial of lyophilized powder with a vial of water. Each CFC has its own device for transfering water to the CFC. Read the instructions enclosed with the product.

When CFC is given the product, batch number and quantity must be recorded in the patient's notes.

When CFC is issued from pharmacy or from the Blood Issue Room fridge (5th floor next to Blood Bank), or the Childrens ED fridge the product, batch number and quantity is recorded at the point of issue.

vCJD
Patients who received British plasma derived clotting factor concentrate or antithrombin concentrate between 1980 and 2001 are designated 'at risk of vCJD for public health purposes'. All such patients have an alert in their medical record. Infection control has a list of identified patients.

If these patients require surgical or endoscopic interventions special measures may need to be taken to decontaminate/quarantine the instruments/scope used. When in doubt, or for emergencies, instruments used must be quarantined until the next working day when a decision about cleaning the instrument and or quarantining can be made.

Previously Untreated Patients
Exposing a Previously Untreated Patient (PUP) to clotting factor is a major decision. Even in the days of recombinant CFCs. Wherever possible recombinant products should be used rather than plasma derived products. There are no known infection risks with recombinant products but the history of haemophilia teaches us caution. This decision should be made by a Consultant Haematologist. PUPs should be consented for concentrate: risks: allergy, inhibitors.
Having said that we must treat the bleed/prevent bleeding effectively.

Prophylaxis.
Clotting factor or Emicizumab is given prophylactically to prevent bleeds.
Newly diagnosed severe haemophilia A and B:
The decision when to start and with what product is made by Nottingham Paediatric Haematologists
Home treatment.
All patients (or their parents) with severe (and some moderates) are trained to administer their own treatment at home. Training done as for prophylaxis. Home treatment is issued by the anticoagulation nurses. See SOP for stock control etc.

Most patient with severe haemophilia A patients are now on emicizumab (a monoclonal antibody which mimics the tenase cofactor activity of VIIIa). Any bleed in a patient on emicizumab must be discussed with a Comprehensive Care Centre. register).

Overall management of inhibitor patients must be discussed with a Comprehensive Care Centre.

Acquired haemophilia
This is a rare autoimmune condition when a person develops antibodies against FVIII. Typically the person is elderly and has other autoimmune conditions or an underlying malignancy. Bleeding can be very severe and include extensive skin bleeding with involvement of the underlying subcutaneous tissue and muscle.
See the acquired haemophilia guideline on KOHA.

Rare bleeding disorders

Fibrinogen deficiency.

For mild bleeding or minor surgery in afibrinogenaemia, hypofibrinogenaemia or haemorrhagic dysfibrinogenaemia, consider tranexamic acid 15–20 mg/kg or 1 g four times daily alone.

For severe bleeding or major surgery in afibrinogenaemia, hypofibrinogenaemia or haemorrhagic dysfibrinogenaemia, consider fibrinogen concentrate 50–100 mg/ kg, with smaller doses repeated if necessary at 2–4 d intervals to maintain fibrinogen activity >1 0 g/l .

Consider pathogen-reduced cryoprecipitate 15–20 ml/kg if fibrinogen concentrate is unavailable.

Factor VII (7) deficiency.

The level of factor 7 needed for surgical haemostasis is 10 – 20%. Cases with a factor 7 > 20% are very unlikely to bleed. People with factor 7 of 40 – 60% are asymptomatic (although the prothrombin time may be prolonged).

Cases with F7D should be identified as at a **higher risk of bleeding if the FVII activity is < 10%,** or if there is another coagulopathy or a personal history of bleeding.

For mild bleeding or minor surgery in higher bleeding risk cases, and for all bleeds and surgery in low bleeding risk cases, consider tranexamic acid 15–20 mg/kg or 1 g four times daily alone.

For severe bleeding or major surgery in higher bleeding risk cases, consider rFVIIa 15–30 mcg/kg repeated if required every 4–6 h, usually for a minimum of three doses.

Factor X (10) deficiency.

For mild bleeding or minor surgery in FXD consider tranexamic acid 15–20 mg/kg or 1 g four times daily alone (2C).

For severe bleeding or major surgery in F10D, consider PCC (i.e. Octaplex) 20–30 (FIX) iu/kg with further PCC 10–20 (FIX) iu/kg at 24-h intervals if required, adjusted to maintain FX activity > 20%.

Factor XI (11) deficiency

Cases with F11D should be identified as at a higher risk of bleeding if the FXI activity is < 10% or if there is another coagulopathy, a personal history of bleeding or if surgery comprises dental extraction or involves the oropharyngeal or genitourinary mucosa.

For minor bleeds or minor surgery in higher bleeding risk cases, and for all bleeds or surgery in low bleeding risk cases, consider tranexamic acid 15–20 mg/kg or 1 g four times daily for 5–7 d.

For severe bleeds or major surgery in high bleeding risk cases, consider a combination of Solvent Detergent-FFP (Octaplas) 15–25 ml/kg and tranexamic acid 15–20 mg/kg or 1 g four times daily.

Factor XII (12) deficiency.
Does not cause bleeding even though it prolongs the APTT.

3. References (including any links to NICE Guidance etc.)

NB: all references are available via the UKHCDO website:

Guidelines for the management of acute joint bleeds and chronic synovitis in haemophilia. A United Kingdom Haemophilia Centre Doctors' Organisation (UKHCDO) guideline. Haemophilia (2017), 1-10.

The diagnosis and management of von Willebrand disease: a United Kingdom Haemophilia Centre Doctors Organization guideline approved by the British Committee for Standards in Haematology. Br J Haematol, 10.1111/bjh.13064 2014.

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

4. Documentation Controls

Reference Number CG-T/2025/175	**Version:** 3		**Status** Final	Dr A McKernan
Version / Amendment History	Version	Date	Author	Reason
	1	2014	Dr A McKernan	New guideline
	2	2019	Dr A McKernan	Review
	3	2024	Dr A McKernan	Review
Intended Recipients: All Haematology Consultants Haematology Specialist Registrar All Haematology CNS's Consultant Surgeon Senior Coagulation BMS Haematology Secretaries				
Training and Dissemination: How will you implement the Clinical Guideline, cascade the information and address training.				
Development of Guideline: Dr A McKernan **Job Title:**Haematology Consultant				
Consultation with: Thrombosis Committee				
Linked Documents: State the name(s) of any other relevant documents				
Keywords:				
Business Unit Sign Off			**Group:**Thrombosis Committee **Date:**2024	
Divisional Sign Off			**Group:**CDCS Division **Date:**Dec 2024	
Date of Upload			Feb 2025	
Review Date			Feb 2028	
Contact for Review			Dr A McKernan	

5. Appendices

OPERATION PLAN TEMPLATE FOR PATIENTS WITH HEREDITARY BLEEDING DISORDERS.

NAME:
HOSPITAL NUMBER:
DATE:

Procedure:
Surgeon:
Anaesthetist:
Anaesthetic:
Haematologist:
Thromboprophylaxis:

Bleeding Disorder:
Factor Level:
FVIII Inhibitor:
Basic clotting: PT, APTT, Fibrinogen, Platelet count.

Virology:
vCJD: Is/is not at risk of vCJD for public health purposes.

Clotting factor concentrate:
Weight :

General principles:

- To elevate the clotting factor to 100% preoperatively and maintain the level between 50 – 100 % for up to 7 days postop.
- Clotting factor concentrate will be given twice daily (approx 10 am and 10 pm) by IV bolus. Unless FIX which is given once daily.
- Blood will be taken for clotting factor levels pre and 20 mins post the morning dose.
- All patients with hepatitis B and/or C and/or HIV will have a full clotting screen (PT, APTT, Fibrinogen, d dimers), FBC and IP with each pre level.
- In addition, **on the day of operation,** a mid pm clotting factor level will be done and an extra dose of clotting factor concentrate given as necessary.
- **ALL BLOODS MUST BE TAKEN TO THE HAEMATOLOGY LAB WITHOUT DELAY AND HANDED TO THE COAGULATION TECHNICIAN WHERE THEY WILL BE DONE URGENTLY.**

Who will dose, give the clotting factor concentrate, and do the bloods?

- Dr Mckernan will dose the clotting factor Mon – Fri. At the weekend the on-call haematology consultant will decide on the dose.
- The morning doses will be given by the haematology CNS
- Clotting factor levels and other bloods will be taken by the haematology CNS.
- The evening doses will be given by the F2/SHO covering the ward. Dr McKernan to liase with the F2/SHOs.

Calculating the dose of clotting factor concentrate.

The dose of FVIII needed to achieve the desired increase in FVIII level is calculated by the following formula:

$$\frac{\text{(Desired rise in FVIII – baseline FVIII) x wt (kg)}}{2} = \text{Units of FVIII}$$

FVIII has a half life of 12 hours (shorter following major surgery) so dosing is BD.

For FIX:
Desired rise in FIX x body wt in Kg = Units of FIX.

The half life of FIX is 18 – 30 hours so repeated dosing is done once daily at a maximum.

DAY OF OPERATION.

Morning (9am): Clotting factor given with pre and post levels.
FACTOR LEVELS MUST BE SATISFACTORY BEFORE PATIENT GOES TO THEATRE.

4 PM: Clotting factor level. Further clotting factor may be given.

Evening (9pm): Clotting factor given.

POSTOPERATIVE DAYS 1 - 7:

9am: Clotting factor level. FBC, full clotting screen, IP.
10 am: Give morning dose and take post FVIII level 20 mins later..

10 pm: Give evening dose.

Following the above a period of prophylaxis may be necessary to cover rehab and physio.