Bleeding Disorders - Surgical Procedures - Full Clinical Guideline

Reference no.: CG-HAEM/2023/011

1. Introduction

Patient with bleeding disorders may require surgery for conditions related to their disorder (eg orthopaedic procedures in people with haemophilia) or unrelated diseases.

Clearly the planning of surgery for patients with bleeding disorders will require additional planning and interaction with other members of the healthcare team than for other patients.

2. Aim and Purpose

To ensure the safe management of surgery for people with bleeding disorders.

3. Main body of Guidelines

Referral for surgical procedures and location of surgery

All surgery in patients registered with the Derby Haemophilia centre must be discussed with the Derby haemophilia team as soon as possible.

Any procedures requiring specialist haemophilia expertise to be available out of hours will be done at Nottingham (or the closest Comprehensive Care Centre eg. Birmingham or Sheffield). In these cases the location of surgery will be decided by the Regional Haemophilia MDT.

All patients having surgery need a surgical plan. A draft plan is shown in Appendix 1. The surgical/anaesthetic teams should be made aware of the plan in advance of the surgery. A copy may be given to the patient.

Elective surgery

1. Logistical considerations:
   Surgery should ideally be scheduled early in the week (Monday – Wednesday) and early in the day (mid – late morning) in order to provide optimal support. Late afternoon scheduling should be avoided where possible.
   The surgical team must liaise with the haemophilia team to ensure that adequate Clinical Nurse Specialist and laboratory support is available to support the surgery. If this is not done, surgery may have to be re-scheduled.
   The on-call haematology team need to be made aware of all post-op patients including plans for monitoring and replacement therapy as appropriate.

2. Investigations prior to surgery
   All patients must have the following investigations clearly documented in the notes and on the surgical plan (Appendix 1)
   a. Up to date weight
   b. Baseline coagulation levels
   c. Inhibitor screen/assay (for patients with haemophilia)
   d. Results of trial of desmopressin (where applicable)

1. Regional anaesthesia
   In preparation for surgery, the planned route of anaesthesia must be clearly documented.
   Regional anaesthesia may be contra-indicated in certain situations where haemostasis cannot be guaranteed eg haemophilia with inhibitors treated with bypassing agents, von Willebrands disease where VWF activity has not been adequately corrected by haemostatic treatment, and bleeding disorders with uncertain bleeding risk.
   A clear plan for the circumstances for regional anaesthesia must be documented on the surgical plan.

2. Haemostatic therapy
   a. The plan for haemostatic therapy must be clearly documented giving name of concentrate (where applicable) and initial dose.
   b. Target pre-operative factor levels should be documented on the surgical plan.
   c. A plan for target levels post-operatively should be documented on the surgical plan.
   d. Recommendations on target factor levels depending on the nature of the surgical procedure can be found in appendix 2.
   e. Adequate stocks of haemostatic therapy must be available in advance of planned surgery. The haemophilia team must ensure that concentrates and other drugs are readily available in clinical areas. Back up stocks will always be available in blood banks.

3. Laboratory monitoring
   a. A clear plan for laboratory monitoring must be documented on the surgical plan.
   b. The haemostasis laboratory should be informed as far in advance of planned surgery as possible to ensure prompt provision of assay results. This will usually be the responsibility of the haemophilia CNSs.
   c. Currently monitoring of replacement therapy in haemophilia A uses the 1 stage F8 assay. Consideration of the use of chromogenic assays for monitoring may be required in specific circumstances eg certain extended half-life F8 products. Any specific monitoring requirements must be discussed with the laboratory well in advance of the surgery and clearly documented.
   d. The likely frequency of laboratory monitoring must be discussed with the laboratory.
   e. Clotting factor replacement and blood tests for monitoring during normal hours is done by the haemostasis CNS. Blood samples must be taken by the CNS by hand to the lab and handed directly to the coagulation lab technician.

4. Thromboprophylaxis
   Venous thrombosis is uncommon in patients with significant bleeding disorders but has been reported in association with coagulation factor replacement (especially when supra-normal levels have been achieved).
   A plan for thromboprophylaxis must be documented. Most patients with haemophilia A will not require pharmacological thromboprophylaxis unless supra-normal levels of F8 have been achieved with haemostatic therapy and surgery is deemed high risk for thrombosis. Consideration of thromboprophylaxis should be made in patients with haemophilia B who achieve normal levels of F9 especially following high risk procedures (eg lower limb or abdomino-pelvic or cancer surgery).
   Mechanical thromboprophylaxis is suggested for all patients undergoing surgical procedures which carry an increased risk of thrombosis.
   Many patients with mild bleeding disorders such as VWD can receive standard pharmacological thromboprophylaxis depending on post-operative factor levels. This should be assessed by the haemophilia team on a case by case basis. Aspirin should be avoided in VWD and platelet function disorders.

A plan for thromboprophylaxis on discharge must be included in the surgical plan.

5. Analgesia
   Post-operative pain should be managed with advice from the pain management team in difficult cases.
   Non-steroidal anti-inflammatory drugs should be avoided in patients with bleeding disorders. This should be clearly documented on the patient's prescription chart.
   Opiate analgesia and paracetamol are safe for use in patients with bleeding disorders.
   Intra-muscular injections are contra-indicated.
   Neuro-axial block (Spinal anaesthesia) may be considered in patients with bleeding disorders in whom haemostasis can be reliably corrected and measured eg haemophilia without inhibitors. Safety needs to be assessed on a case by case basis, and factor levels must be clearly documented in the patient's surgical plan. If neuro-axial block is necessary spinal anaesthesia is acceptable after a bolus of clotting factor. Epidural anaesthesia should not be used unless there are compelling reasons why (in which case there must be a clear plan for catheter removal) .Neuro-axial blocks are contra-indicated in disorders where haemostasis cannot be guaranteed/monitored appropriately.
6. Physiotherapy
   A plan for factor replacement therapy should be documented for patients with severe bleeding disorders who require post-operative physiotherapy eg following joint replacement surgery.

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7. 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 sticker in the front of their notes. Infection control have 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. For further advice go to the trust policy at https://derby.koha-ptfs.co.uk/cgi-bin/koha/tracklinks.pl?uri=https://derby.koha-ptfs.co.uk/cgi-bin/koha/opac-retrieve-file.pl?id=c608655143f27e616b6e0eaf90b017b7;biblionumber=1759 and contact infection control

8. Discharge planning
   The surgical plan should include plans for continuation of factor treatment post-discharge where applicable.
   Plans for suture removal should also be documented where applicable.
   If pharmacological thromboprophylaxis is to be given, a clear plan must be in place on discharge for dose and duration.
   Patients and carers must have clear information on discharge of how to contact the haemophilia team in the event of post-operative bleeding complications.

Emergency surgery

All patients with bleeding disorders who require emergency surgery must be discussed with the haematology consultant on call.
Haemostatic replacement therapy must be given and monitored in accordance with guidance for elective surgery.
See appendix 2 for guidance on target factor levels.

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

5. Documentation Controls

Development of Guideline:	Angela McKernan
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Approved By:	Thrombosis group 5/3/19 Reviewed A. McKernan - Dec 2023 CDCS Division - Dec 2023
Review Date:	Dec 2026
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6. Appendices

Appendix 1.

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 60 – 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 every 18 – 24 hours.
- For joint replacement factor will be given by continuous infusion for 3 – 4 days to maintain the level between 80 – 120%; then BD bolus.
- 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,** an end of procedure/ 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.

DAY OF OPERATION.

Morning (9am):	Clotting factor given with pre and post levels. Factor levels must be satisfactory before patient goes to theatre**.** **If there is no inhibitor and the response to CFC is known surgery may proceed without waiting for the factor levels.**
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.

Dr A McKernan.
Consultant Haematologist.

Circulation:
All Haematology Consultants
Haematology Specialist Registrar
All Haematology CNS's
Consultant Surgeon
Senior Coagulation BMS
Haematology Secretaries

Appendix 2

Suggested plasma factor levels and duration of administration

	Haemophilia A		Haemophilia B	
	Desired factor level (iu/dL)	Duration (days)	Desired factor level (iu/dL)	Duration (days)
Joint replacement	80 – 120 By continuous infusion	4 Then as for major surgery	80 – 120 By continuous infusion	4 Then as for major surgery
Major surgery Pre-op	80-100		60-80	
Post-op	60-80 40-60 30-50	1-3 4-6 7-14	40-60 30-50 20-40	1-3 4-6 7-14
Minor Surgery Pre-op	50-80		50-80	
Post-op	30-80	1-5 depending on procedure	30-80	1-5 depending on procedure