

1. Introduction and Who Guideline applies to

This guideline is for medical teams caring for patients with bleeding disorders who are due for an intervention or surgical procedure.

2. Guideline Standards and Procedures

The management of surgical and dental procedures for people with bleeding disorders must be approached with care and should involve multi-disciplinary team (MDT) discussion and excellent communication between those involved. Typically the MDT would include: operator, operator nursing team lead, anaesthetist, haemostasis team, laboratory and patient. The patient's GP should be kept informed at all times.

In order to successfully manage haemostasis in people with bleeding disorders, the following should be identified and assessed:

- Date/time/location of procedure
- Bleed risks associated with the procedure; ideally as assessed by the operator but see also Appendix 1 for bleed risk assessment for surgical procedures and Appendix 2 for dental procedures.
- Patient's bleed risk (i.e. severity of diagnosis of bleeding disorder)
- Details of the recovery period- time required for healing and/ or rehabilitation. This includes length of in-patient stay, patient support at home, patient ability to self-treat with IV injections if necessary

Once planned, the MDT should follow the procedure below:

1. Surgical/dental team to inform haemostasis team of the procedure. Required details include: name of procedure, planned time and date, location, mode of anaesthesia, bleeding risk of the procedure and recovery time.
2. Haemostasis team to arrange for a written plan for peri-operative haemostasis.
3. Haemostasis team to discuss and sign off written plan at haemostasis MDT meeting (or sooner depending on urgency); then send plan to surgical team, haemostasis MDT, patient, +/- laboratory (if specialist tests required)

The haemostasis team will be responsible for ensuring action points from the peri-operative plan. This might include: supply of factor to ward nursing teams, education regarding factor administration, coordinating post-procedural blood samples and their delivery to the special haematology laboratory, consideration of venous access and post procedural home treatments.

The written plan devised by Haemophilia team should contain information with specific instructions about timing, dose of factor concentrate and laboratory tests required.

Important considerations for the haemostasis team:

- Have inhibitors or anti-platelet antibodies been considered and excluded pre-operatively?

- Day/time of procedure: earlier in the day and earlier in the week are preferable when considering staff and laboratory availability in the post-procedure setting.
- Is there sufficient availability of haemostatic agent required for this patient?
- How and where will haemostatic agents be given in the early post-operative period? Can these be given at home with the help of an indwelling venous catheter for example?

Specific therapies, dosing and timing are beyond the scope of this guideline and management plans are to be highly individualised in order to mitigate risks.

DDAVP may be useful haemostatic treatment for minor bleeding and invasive procedures in responsive patients with mild Haemophilia A, some types of Von Willebrand disease and some platelet function disorders (without medical contraindication)- see Desmopressin guideline for more details.

Tranexamic acid, an antifibrinolytic can be used alone or in conjunction with other perioperative haemostatic treatments depending on bleeding risk.

Anaesthesia: For patients undergoing surgery avoid neuraxial anaesthesia. If required, it should be performed under adequate clotting factor coverage as safety of neuraxial procedures has not been established in patients with haemophilia.

Thromboprophylaxis: For patients with bleeding disorder undergoing surgery, routine thromboprophylaxis is avoided unless advised by haemophilia team.

3. Education and Training

Nil

4. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Review of procedural success	Adherence to plan; bleed rates	Haemophilia MDT	3m	MDT minutes

5. Supporting References (maximum of 3)

- 1. WFH Guidelines for the Management of Hemophilia, 3rd edition:** Alok Srivastava, Elena Santagostino, Alison Dougall, Steve Kitchen, Megan Sutherland, Steven W. Pipe, Manuel Carcao, Johnny Mahlangu, Margaret V. Ragni, Jerzy Windyga, Adolfo Llinás, Nicholas J. Goddard, Richa Mohan, Pradeep M. Poonnoose, Brian M. Feldman, Sandra Zelman Lewis, H. Marijke van den Berg, Glenn F. Pierce, on behalf of the WFH Guidelines for the Management of Hemophilia panelists and co-author

6. Key Words

Haemophilia, von willebrand disease, platelet function disorders, bleeding disorder, perioperative management, clotting factor, coagulation factor, factor deficiency

CONTACT AND REVIEW DETAILS	
Guideline Lead (Name and Title) and reviewed by Dr Sandhya Munireddy and Dr Styliani Salta Consultant in Haemostasis and Thrombosis	Executive Lead
Details of Changes made during review: 1. Section on DDAVP, tranexamic acid, anaesthesia and thromboprophylaxis included. 2. Generic template removed	

Appendix 1. Procedural bleed risks when considering management of bleeding disorders

High bleed risks: requiring maintenance of higher factor levels for a longer duration.

Major surgery: penetration/exposure of a body cavity, extensive tissue resection.

All major surgeries

All vascular surgeries

All cardiac/ cardiothoracic surgeries

Cardiovascular interventions

Pacemaker or defibrillator placement

Coronary intervention and angiography

Electrophysiological testing/ablation

Ophthalmologic surgeries

Peri-orbital surgery

Vitreoretinal surgery

ENT surgeries

Sinus surgeries

Biopsy or removal of nasal polyps

Thyroidectomy

Parotidectomy

Septoplasty

Turbinate cautery

Dental(see also Appendix 2)

Reconstructive procedures

Orthopaedic surgeries

Arthroplasty

Arthroscopy

Joint replacement surgery- Shoulder/ foot/hand surgeries

Spinal surgery

Gynaecologic surgery

Hysterectomy

Bilateral tubal ligation

Laparoscopic surgery

Cancer surgery

General surgery

Surgery on spleen/liver/kidney

Bowel resection

Laparoscopy

Abdominal hernia surgery

Laparoscopic Cholecystectomy

Lymph node biopsy

Haemorrhoidectomy

Gastroenterology

- Polypectomy
- Percutaneous endoscopic gastrostomy
- Percutaneous liver biopsy
- Endoscopic Ultrasound with fine needle biopsy
- Endoscopic biliary or pancreatic sphincterotomy
- Variceal banding

Urology and renal

- TURP
- Bladder tumour resection
- Renal biopsy
- Extracorporeal shock wave lithotripsy

Neurosurgery/neuraxial procedures

All procedures including lumbar puncture and myelography

Interventional radiology

- Percutaneous trans hepatic cholangiography or nephrostomy
- Percutaneous drainage of liver abscess or gall bladder
- Organ biopsy
- Hickman and tunnelled dialysis catheter placement

Respiratory

- Chest tube placement

Low bleed risk: may require haemostatic intervention but at a lower intensity and for a shorter period of time

- Minor surgery eg skin
- Endoscopies without biopsy

Minor dermatological procedures : Excision of Basal and Squamous cell skin cancers /actinic keratosis and premalignant / malignant lesions

Dental **

- Dental fillings/ cleaning
- Simple dental extraction
- Restoration prosthetics
- Endodontics

Ophthalmology

- Cataract extraction

Gastroenterology

- Diagnostic endoscopy with or without biopsy
- ERCP without sphincterotomy

Urology

Cystoscopy without biopsy

ENT

Diagnostic fiberoptic laryngoscopy or nasopharyngoscopy

Fine needle aspirate

Vocal cord injection

Gynaecological procedures

Diagnostic hysteroscopy

Colposcopy

Insertion of intrauterine devices

Appendix 2. Bleed risks and management suggestions for dental procedures in the context of haemophilia and bleeding disorders

Procedures typically requiring factor replacement/haemostatic support. Consider additional tranexamic acid.	Procedures typically not requiring specific haemostatic support. Consider tranexamic acid.
Inferior dental block	Buccal infiltration
Lingual infiltration	Intra-papillary injection
	Intra-ligamentary injection
Dental extraction(s)	
Implants/complex procedures	Scaling and polishing
	Orthodontic assessment/fitting
	Root canal