

Consensus statement by hospital based dentists providing dental treatment for patients with inherited bleeding disorders*

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ABSTRACT

Avoidance of dental care and neglect of oral health may occur in patients with inherited bleeding disorders because of concerns about perioperative and postoperative bleeding, but this is likely to result in the need for crisis care, and more complex and high-risk procedures. Most routine dental care in this special needs group can be safely managed in the general dental setting following consultation with the patient's haematologist and adherence to simple protocols. Many of the current protocols for dental treatment of patients with inherited bleeding disorders were devised many years ago and now need revision. There is increasing evidence that the amount of factor cover previously recommended for dental procedures can now be safely reduced or may no longer be required in many cases. There is still a need for close cooperation and discussion between the patient's haematologist and dental surgeon before any invasive treatment is performed. A group of hospital based dentists from centres where patients with inherited bleeding disorders are treated met and, after discussions, a management protocol for dental treatment was formulated.

Keywords: Blood coagulation disorders/complications, dental care, haemophilia, haemostatics/therapeutic use, von Willebrand disease.

Abbreviations and acronyms: HDS = hospital dental service; HTC = haemophilia treatment centre; NSAIDS = Non-steroidal anti-inflammatory drugs.

(Accepted for publication 26 September 2010.)

BACKGROUND AND AIM

Dental treatment protocols for the management of patients with inherited bleeding disorders vary between Australian states and territories (Table 1).^{1,2} Many of these protocols are based on the Stubbs and Lloyd article published in 2001,³ and recommendations by the Australian Health Minister Advisory Council FVIII and FIX guidelines 2006,⁴ but vary in their complexity and content. This consensus statement was developed in

order to provide up-to-date advice regarding the bleeding risk and the possible need for systemic haemostatic support (e.g. replacement factor concentrates, desmopressin, etc.) during the dental treatment of patients with inherited bleeding disorders. It does not provide detailed protocols for dental procedures for these patients.

The statement was developed following a meeting in October 2009 and extensive consultation between the authors. The final document represents a consensus view of the listed authors. The available literature and international guidelines were reviewed. However, there is little evidence based data for many aspects of dental

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Table 1. Common inherited bleeding disorders^{1,2}

	Factor affected	Bleeding severity and factor levels
Haemophilia A ¹	Factor VIII levels decreased	Severe: <1 IU/dL Moderate: 1–5 IU/dL Mild : >5–<40 IU/dL
Haemophilia B ¹	Factor IX levels decreased	Severe: <1 IU/dL Moderate: 1–5 IU/dL Mild : >5–<40 IU/dL
von Willebrand disease ²		
Type 1	VWF:Ag levels decreased	Variable, usually mild-to-moderate
Type 2	Dysfunctional VWF	Variable, usually moderate
Type 3	VWF absent	Severe (VWF:Ag undetectable, factor VIII <10 IU/dL)

VWF:Ag = von Willebrand factor antigen.

care for patients with inherited bleeding disorders. Therefore, some recommendations are by necessity based on clinical experience and professional opinion.

Overview of dental care in patients with inherited bleeding disorders

Dental procedures, such as extractions and periodontal surgery, are associated with postoperative bleeding, which is generally self-limiting. However, in patients with an inherited bleeding disorder (Table 1), relatively minor invasive procedures may precipitate prolonged bleeding. Excessive bleeding is distressing for both patients and clinicians, and can delay completion of the procedure, compromise wound healing and predispose to infection. The bleeding risk associated with dental procedures varies with how easy it is to access the site and apply local haemostatic measures. For a simple extraction, local haemostatic measures, such as pressure or topical agents can usually be applied to the potential site(s) of haemorrhage. In contrast, there may be little or no access to bleeding sites following deep spatial or cavity surgery and some flap surgery.⁵ In general, superficial and visible bleeding is easier to manage than deep invisible bleeding.

Some minor procedures with a low risk of bleeding can be performed without systemic haemostatic therapy. However, for higher-risk procedures (extractions and oral surgery), appropriate systemic haemostatic cover may need to be considered and should be managed in consultation with a haemophilia treatment centre (HTC) or haematologist. These decisions must take into account not only the nature and severity of the patient's bleeding risk but also the type, location and extent of the dental procedure, and the expertise and experience of the treating dentist.

Haemostatic protocols may use systemic therapies (when needed), together with suturing and additional local measures, such as Surgicel[®] (Johnson and Johnson, UK), Gelfoam[®] (Pharmacia and Upjohn, Michigan, USA), cyanoacrylate tissue adhesives, fibrin

glue and surgical splints, in conjunction with perioperative antifibrinolytic agents, such as tranexamic acid mouthwash.⁵ These local techniques may enable oral surgery procedures to be undertaken without the need for any additional systemic haemostatic cover.^{6,7}

Oral healthcare in patients with inherited bleeding disorders should have a strong preventive focus. The prevention of dental disease, which is a vital component of oral care for these patients, has been adequately covered elsewhere,^{6,8} so is not discussed in this statement. Adequate preventive measures and regular dental review examinations can reduce the need for remedial interventions and emergency procedures.

Responsibility for dental care

Dentists involved in the care of patients with inherited bleeding disorders need to develop an understanding of the nature of the condition, the severity of the disease and the previous response to treatment in order to plan safe and effective dental treatment.

Ideally, the dental care requirements for all patients with inherited bleeding disorders should be assessed initially in a hospital dental service (HDS) in consultation with a referring HTC. A HDS in conjunction with a HTC should continue to manage the dental care of patients with a severe bleeding tendency. However, ongoing dental care for most patients with either a moderate or mild inherited bleeding disorder can be managed in the general dental practice environment.

Some patients with severe inherited bleeding disorders may be on long-term prophylactic factor replacement to minimize complications such as haemarthrosis, and may require little or no adjustment to their factor replacement. Patients with less severe inherited bleeding disorders may require haemostatic support to be coordinated with their dental treatment. Haemostatic support is not without risk to the patient, and so must be carefully considered in the context of the dental treatment required. Furthermore, the cost of factor replacement may easily exceed the cost of the proposed dental treatment. Consideration should be given to the

scheduling and coordination of dental procedures to minimize the need for factor replacement.

Private general dental or community dental practitioners may perform routine non-invasive dental care in consultation with the patient's haematologist and HTC dentist. Specific dental advice can be sought from hospital dentists associated with regional HTC. For children or adults with extensive oral disease, where multiple appointments may be required, comprehensive oral treatment under general anaesthetic in consultation with a HTC should be considered.

TREATMENT CONSIDERATIONS

Local anaesthesia and pain control

The use of local anaesthetics in patients with bleeding disorders should be discussed with a HTC or haematologist. Even submucosal infiltrations have the potential to cause haematomas in patients with severe bleeding disorders, although the risk is extremely low with modern single-use needles.

Oral injections of local anaesthetic pose varying degrees of risk for patients with inherited bleeding disorders. Typically, infiltrations can be used without systemic haemostatic cover.^{6,9} Slow injection will allow time for the local anaesthetic solution to diffuse through the tissues and minimize bruising. A local anaesthetic with a vasoconstrictor should be used where possible because these provide additional local haemostasis.^{6,10} Also, consideration should be given to the use of nitrous oxide-oxygen, and anxiolytic agents to assist with pain control when necessary.¹⁰

In patients with severe bleeding disorders, the use of regional nerve blocks, lingual infiltration and floor-of-mouth injections should be discussed with a HTC/haematologist. In the past, bleeding following deep nerve-block injections (inferior alveolar and posterior superior alveolar) has been implicated in airway obstruction due to haematoma formation in the retromolar or pterygoid space;^{6,10} inferior alveolar nerve blocks have been associated with a greater risk. However, the risk of haematoma is now thought to be low with modern fine-gauge single-use needles. The Gow-Gates technique may be preferable to conventional inferior alveolar nerve block.⁵ Nevertheless when possible, an alternative to nerve blocks, such as intraligamentous or intrapapillary injections should be used;^{6,10} this is strongly recommended in paediatric dentistry. The use of articaine may allow infiltrations to be used for lower molars.⁹ When a nerve block is used, the haemostatic cover requirements may need to be considered, although a recent study found that additional factor support may not be required.⁷ Many severe haemophilia and Type 3 von Willebrand patients are taking ongoing prophylactic factor cover,

which minimizes the risk of bleeding events for dental treatment.

Post-procedure dental pain can usually be controlled with a minor analgesic, such as paracetamol, possibly in conjunction with codeine. Non-steroidal anti-inflammatory drugs (NSAIDs), including aspirin, can adversely affect platelet aggregation. Aspirin should not be used, and the use of other NSAIDs should be discussed with the patient's haematologist.⁶ When appropriate, a long-acting local anaesthetic, such as bupivacaine hydrochloride (MarcaineTM, AstraZeneca, Australia) 0.5% with 1:200 000 adrenaline, is a good option for postoperative analgesia. Long-acting local anaesthetics can give 8 to 10 hours of effective pain control. Patients, and particularly children (and their parents), should be warned of the risk of inadvertent postoperative self-trauma, such as biting their lip, cheek or tongue while the oral soft tissues are numb.

General measures to reduce trauma

When undertaking any procedure in the mouth, damage to the oral mucosa should be minimized. Local trauma can be reduced during routine dental procedures with the careful use of saliva ejectors, exercising care when taking impressions and placing of radiographic films, particularly in the sublingual region. Rubber dams and other techniques should be used to protect soft tissue during restorative procedures.⁶ During oral surgery, surgical techniques should be modified to minimize both intraoperative and postoperative bleeding.⁷

TREATMENTS AND PROCEDURES

Periodontal treatment

Healthy periodontal tissues do not bleed and maintain the dentition so that preventable tooth loss can be avoided. For patients with inherited bleeding disorders who have poor oral hygiene, an immediate treatment plan should be formulated to prevent further damage to the periodontal tissues.⁶ Patients with gingivitis are often reluctant to brush their teeth because of concerns about bleeding gingival tissues, and this can lead to further deterioration of their periodontal health.

For patients with mild periodontal disease, routine supragingival scaling and prophylaxis is unlikely to cause prolonged bleeding; there is usually no specific interventions required to reduce bleeding.⁹ If the gingival condition is poor, or the patient has a moderate or severe bleeding tendency, a 5% tranexamic acid mouthwash (one 500 mg tranexamic acid tablet dissolved in 10 ml of water) may be required to control bleeding following supragingival scaling and cleaning. A HTC/haematologist should be consulted.

Management of patients with severe periodontal disease should be individualized and undertaken by a HDS in conjunction with a HTC/haematologist. Usually, periodontal probing, supragingival scaling and prophylaxis can be done without risk of significant bleeding. Ultrasonic instrumentation may result in less tissue trauma than hand scalers used subgingivally.¹⁰ For severely inflamed tissues, chlorhexidine mouthwashes and gross debridement are recommended to reduce tissue inflammation prior to subgingival scaling.¹⁰ Post-treatment, 5% tranexamic acid mouthwashes are usually effective in controlling protracted bleeding.¹⁰ Treatment may need to be completed over several visits to prevent excessive bleeding.⁶

Periodontal surgery in patients with inherited bleeding disorders is a high-risk procedure with significant risk of bleeding and should only be considered where conservative treatment measures have failed. As periodontal surgery can be a greater challenge to haemostasis than a simple extraction, periodontal surgery must be carefully planned with the need for systemic haemostatic cover determined in consultation with a HTC/haematologist. The risks should be fully explained to the patient.⁶

Extractions and oral surgery

Surgical treatment, including routine dental extractions, for patients with an inherited bleeding disorder must be planned to minimize the risk of bleeding, excessive bruising or haematoma formation. These patients may require additional systemic haemostatic cover.⁶ Treatment protocols, including the degree of factor cover if required, should be determined in close consultation between a dentist with experience in this field and the patient's haematologist. Ideally, a HDS dentist in consultation with a HTC/haematologist should undertake these procedures.

Prior to extraction, efforts should be made to reduce the risk of local infection and inflammation by using topical antiseptics (chlorhexidine or povidone iodine), or antibiotics⁶ if the infection is considered to require more than topical measures. Amoxicillin 500 mg three times a day for seven days is usually appropriate. Construction of a surgical stent may help to protect the surgical site during healing. Surgical techniques should be modified to minimize both intraoperative and postoperative haemorrhage. These measures include: limiting trauma by elective sectioning of teeth; limiting the number of teeth to be removed at a time; reducing the size of flaps; choosing surgical and closure techniques that permit easy access for packing; suturing and cautery; striving to obtain primary surgical closure; and removing all granulation tissue from areas of chronic inflammation.⁵ Although careful preoperative planning will prevent many postoperative problems, post-extraction bleeding may occasionally occur. If this

is the case, consult the HTC/haematologist and consider using additional systemic haemostatic therapy.

Routine normal or assisted exfoliation of primary teeth does not require haemostatic cover. Persistent oozing and bleeding following the procedure should initially be managed with local measures, such as pressure and 5% tranexamic acid mouthwash.

Restorative procedures

Routine restorative dental treatment, including crowns and bridges, is associated with a low risk of bleeding and can be carried out in general dental practice, provided guidelines for local anaesthetic use are followed.^{6,8,9} Consideration should be given to the use of appropriate minimal intervention techniques where indicated. Alternative instrumentation, such as air abrasion and hard tissue lasers may further reduce the need for local analgesia.

Orthodontic treatment

Conventional orthodontic treatment including fixed and removable orthodontic appliances may be used in patients with inherited bleeding disorders along with regular preventive advice and hygiene therapy.⁶ Routine non-extraction orthodontic treatment would not normally require consultation with a HTC or haematologist. Special care should be taken to ensure that the gingiva is not damaged when fitting the appliance and to avoid sharp wires or edges that may traumatize mucosa or gingival. Soft periphery wax can also be used to reduce tissue trauma after appliance insertion. Extractions and minor surgical procedures need to be considered carefully, but can be completed with the precautions already described. Orthognathic surgery is not absolutely contraindicated, but has an unfavourable risk:benefit ratio. The treating practitioner should liaise with a HTC or haematologist during the treatment planning phase if any surgical procedures are being considered to support the orthodontic treatment.

Dentures

There is no contraindication to the use of full or partial dentures.⁶ Prosthetics are unlikely to cause problems provided care is taken to avoid tissue trauma during the fabrication process and the dentures are adjusted to avoid any over extension.

Implant-supported prosthesis

There is little information about the use of implants in patients with inherited bleeding disorders.¹¹ It is reasonable to consider that routine placement of implants would pose no more risk than extraction of

third molars. However, complications such as perforation of the lingual surface of the mandible has the potential to cause deep bleeding that could be difficult to recognize and control. If it is thought that the lingual plate has been perforated, the patient should immediately be referred to a HTC for assessment and treatment if required. Adjunctive surgery, such as bone grafting and sinus lift surgery, are contraindicated. The use of three-dimensional imaging and treatment planning software may help to verify the suitability of a proposed implant site. Until definitive evidence based protocols are established and verified for the use of implants in patients with inherited bleeding disorders, it would be prudent to discuss the proposed use of implants with a HTC or HDS clinician at the treatment planning stage.

Endodontic treatment

Endodontic treatment is a low-risk procedure in patients with an inherited bleeding disorder.⁶ Pulpectomy and pulpotomy procedures can be performed routinely and are generally preferable to extraction. It is important that a pulpectomy is carried out carefully within the working length of the root canal to ensure that the instruments do not pass through the apex of the root canal.⁶ Bleeding in the root canal of a tooth with a closed apex suggests that there is pulp tissue remaining in the canal,⁶ or the apex has been perforated. In teeth with open apices, there is an increased risk of intracanal bleeding. Bleeding can usually be controlled with 4% sodium hypochlorite irrigation, and calcium hydroxide dressing of the root canal.⁶

Pulpotomy can be carried out in primary teeth according to accepted indications and contraindications. Electrosurgical coagulation of the pulp stumps or the use of ferric sulphate may assist with haemostasis.

CONCLUSIONS

Oral healthcare may often be neglected in people with inherited bleeding disorders. Early and ongoing preventive dental care is essential in this patient group to avoid invasive procedures at a later date. This article aims to facilitate not only the delivery of routine dental care in this patient group by encouraging collaboration between the patient's dental clinician and haematologist but also by providing up-to-date information about the bleeding risk associated with varying dental operative procedures.

ACKNOWLEDGEMENTS

The authors declare that there are no relevant conflicts of interest. The AHCDO receives funding from the National Blood Authority for administrative purposes.

TREATMENT MANAGEMENT RECOMMENDATIONS

Haemophilia – severe: <1% Factor VIII or IX

- Send to HTC for any invasive dental procedures
- Routine scaling and cleaning with 5% tranexamic acid mouthwash 10 ml every 6 to 8 hours for 3 to 5 days unless severe periodontal disease is present, then discuss with HTC or HDS clinician
- Routine endodontic, restorations and orthodontic treatment with caution
- IDN block poses some risk of haematoma, discuss with HTC or HDS clinician

Haemophilia – moderate: 1%–5% Factor VIII or IX

- Send to HTC for any oral surgery or ring HTC dentist for advice
- Routine scaling and cleaning with 5% tranexamic acid mouthwash 10 ml every 6 to 8 hours for 3 to 5 days unless severe periodontal disease, then HTC or HDS clinician
- Routine endodontic, restorations and orthodontic treatment do not require any cover
- IDN block poses a lower risk of haematoma

Haemophilia – mild: >5% Factor VIII or IX

- Oral surgery careful extraction, 5% tranexamic acid and SURGICEL (Johnson & Johnson, New Yorkshire, UK) in socket, suture socket tightly with 4-0 MONOCRYL (Ethicon, Johnson & Johnson, Somerville, New Jersey, USA) or 4-0 VICRYL (Ethicon, Johnson & Johnson, Somerville, New Jersey, USA) sutures, 5% tranexamic acid mouthwash 10 ml every 6 to 8 hours for 3 to 5 days postoperative
- Scaling and prophylaxis with 5% tranexamic acid mouthwash if post-cleaning oozing a problem
- Routine endodontic, restorations and orthodontic treatment do not require any cover
- IDN block poses a low risk of haematoma

von Willebrand's Disease – Type 1: (60–80%)

Quantitative defect

- Oral surgery careful extraction, 5% tranexamic acid and SURGICEL (Johnson & Johnson, New Yorkshire, UK) in socket, suture tightly with 4-0 MONOCRYL (Ethicon, Johnson & Johnson, Somerville, New Jersey, USA) or 4-0 VICRYL (Ethicon, Johnson & Johnson, Somerville, New Jersey, USA) sutures
- Scaling and cleaning with 5% tranexamic acid mouthwash 10 ml every 6 to 8 hours for 3 to 5 days unless severe periodontal disease, then discuss with a HTC or HDS clinician

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- Routine endodontic, restorations and orthodontic treatment do not require any cover
- IDN block low risk of haematoma

**von Willebrand's Disease – Type 2A and 2B: (20–30%)
Qualitative defect**

- Oral surgery careful extraction, 5% tranexamic acid and SURGICEL (Johnson & Johnson, New Yorkshire, UK) in socket, suture tightly with 4-0 MONOCRYL (Ethicon, Johnson & Johnson, Somerville, New Jersey, USA) or 4-0 VICRYL (Ethicon, Johnson & Johnson, Somerville, New Jersey, USA) sutures
- Scaling and cleaning with 5% tranexamic acid mouthwash 10 ml every 6 to 8 hours for 3 to 5 days unless severe periodontal disease, then discuss with a HTC or HDS clinician
- Routine endodontic, restorations and orthodontic treatment do not require any cover
- IDN block low risk of haematoma

**von Willebrand's Disease – Type 3: Severe form:
Platelet and Factor VIII**

- Oral surgery: send to HTC, significant risk of mucosal bleeding
- Scaling and cleaning send to HTC or HDS
- Routine endodontic, restorations and orthodontic treatment do not require any cover
- IDN block poses a higher risk of haematoma discuss with HTC or HDS clinician.

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