DEPARTMENT OF HAEMATOLOGY

HAEMOPHILIA CLINIC

Enquiries to: CNS Jane Thomas *Tel:* GRO-C *ext:* GRO-C *Bleep No:* GRO-C *Fax No:* GRO-C

Dr. S. Al-Ismail FRCP FRCPath Consultant Haematologist	Dr. A. Benton FRCP FRCPath Consultant Haematologist	Dr. A. Salamat MRCP MRCPath Consultant Haematologist	Dr. H. Sati MD., MRCP(I) FRCPath Consultant Haematologist	Dr. U. Mohite Consultant Haematologist		Dr. V. Devalier FRCP FRCPath Consultant Haematologist

HN:	**256155**	
Name:		
Address:		**Signed: Dr S Al-Ismail** **Title: Consultant Haematologist** **Date: 21/04/2011**
DOB:		

DIAGNOSIS: **Haemophilia A**

Date: 21/04/2011 **WEIGHT:** **(need repeat weight in 6 months)**	**80 Kg** Dose **must** be recalculated if it has changed by > 10%
FACTOR VIII LEVEL:	**< 1%**
MANAGEMENT:	**If replacement therapy is needed please contact the Consultant Haematologist on call and use:-** **Refacto AF** **For bleeding in joints :- 2500 units** **For bleeds in muscles:- 3000 units** **For life threatening bleeds (e.g. cerebral, tongue) : 4000 units**

Care Plan for Children at School with Haemophilia

NAME:

DOB:

DIAGNOSIS:

SCHOOL:

DATE:

Contact Details:

Family Contact 1
Name:

Relationship:

Number: Home:

Mobile: (1st contact)

Family Contact 2
Name:

Relationship:

Number: Home:

Mobile:

GP:
Name:

Number:

Swansea Haemophilia Centre
CNS Haemophilia Kerry Bowen
or Anne Jones
GRO-C

Hospital Consultant: Dr S Al -Ismail

What is Haemophilia?
When a person's blood cannot clot properly the condition may be known as Haemophilia. Although Haemophilia is a lifelong disorder, it can be managed as part of a normal active life.

CARE REQUIREMENTS:
Please do not hesitate to contact the Haemophilia Centre in Swansea @ Singleton Hospital

1. **Head Injuries**

   - **Any head injury may be serious due to the possibility of internal bleeding .**
   - The child needs to be observed for vomiting, headache , dizziness, drowsiness, swelling/ bruising ,loss of consciousness.
   - If the child has had a head injury the parent should be called- the child should be taken to The Paediatric Assessment Unit (PAU) in Morriston Hospital . The Haemophilia Centre ,Singleton should be contacted

   In addition to above normal first aid measures should be taken.

2. **Any other bumps/injuries/grazes**

   - Minor injuries can be dealt with, with normal first aid measures initially. It is really important that parents are informed about any knocks or bumps as they are most expert at deciding if something needs further action.

   - Mouth or tongue bleeds can be hard to deal with and may need treating by the haemophilia centre. (Sucking on an ice cube may help.)

   - If you have any concern about an injury then contact the parent and/or the haemophilia centre for advice.

3. **Nose Bleeds**

- Normal first aid measures to be used, see guidance given to school first aider . However if the nose bleed exceeds 20 minutes you would be advised to contact the parent /and/or the haemophilia centre.

- A tablet or syrup of transexamic acid can be taken to help if he has had an increasing number of nose bleeds. This would be taken as a preventative measure and be ineffective in controlling a nose bleed at the time of the bleed.The parent can advise when the child needs to take these tablets or syrup.

4. **In Conclusion**

- **If in doubt contact the parent and/or the Haemophilia centre**
- **Minor Injuries can be dealt with ,with normal first aid measures**
- **Head injuries need special attention, please refer to the Haemophilia Centre and Morriston Hospital Paediatric Unit.**

In case of emergency you will need to bring the child to hospital. Instruct the ambulance crew to bring him directly to Morriston Hospital and to contact the Paediatric Department.

Please inform Morriston beforehand and also inform the Haemophilia Department

Updated KB July 2017

GIG CYMRU NHS WALES
Bwrdd Iechyd Prifysgol
Abertawe Bro Morgannwg
University Health Board

Singleton Hospital
Sketty Lane
Swansea
SA2 8QA
Tel: 01792 205666

Peri- Operative Plan for Patients with Bleeding Disorders

Haemophilia Centre Tel: 01792 200368 Fax: 01792 200368

Name: Hospital Number: NHS Number: D.O.B: **Weight**:	**Location** for planned admission/procedure: Cons. Haematologist: Specialist Cons:

Diagnosis:
Baseline Levels:
Inhibitor: No
Usual Product:
Co-morbidities:

Additional comments/considerations:
Name:

Planned Procedure:
Date & **Time** of Procedure: Date of Admission:
Anticipated Length of Stay:

All patients are recommended for thromboprophylaxis in line with procedure, weight, creatinine clearance as standard **unless** specified here:

Treatment Plan

If you have any concerns about Haemostasis then please contact staff from the Haemophilia centre on Ext: 7768.

Factor concentrates will be provided and prescribed by SHC. If post-procedure review is specified as "required" in the plan, please do not discharge the patient home before Haemophilia team review.

Enquiries: 9am-5pm SHC: GRO-C SpR Bleep: GRO-C
Emergency Haematology advice is available OOH, ask for Haematology Consultant on call

GIG CYMRU NHS WALES
Bwrdd Iechyd Prifysgol
Abertawe Bro Morgannwg
University Health Board

Singleton Hospital
Sketty Lane
Swansea
SA2 8QA
Tel: 01792 205666

Peri- Operative Plan for Patients with Bleeding Disorders

Haemophilia Centre Tel: 01792 200368 Fax: 01792 200368

Name: XXXXXXXXXXX Hospital Number: XXXXXXXXXXXX NHS Number: XXXXXXXXXXXXX D.O.B: XXXXXXXXXXXXXX **Weight**: 49.5 kgs	**Location** for planned admission/procedure: Singleton Cons. Haematologist: Dr. S. Al-Ismail Specialist Cons: Dr. S.T.Browning

Diagnosis: Haemophilia A -Severe
Baseline Levels: < 1 iu/dL
Inhibitor: No
Usual Product: Refacto AF
Co-morbidities:

Additional comments/considerations:

- Aim of replacement therapy with Factor VIII (Refacto AF) is to keep post infusion level at 100iu/dl and pre infusion level above 50iu/dl.
- Please inform coagulation lab in Morriston (e-mail management plan)

Planned Procedure: Mastoid Exploration
Date & **Time** of Procedure: Tuesday 23.05.17 Date of Admission: 23.05.17
Anticipated Length of Stay:

All patients are recommended for thromboprophylaxis in line with procedure, weight, creatinine clearance as standard **unless** specified here:

Treatment Plan

- **Start on Tranexamic acid 1gm TDS on 22 May and continue for 14days post-operative**
- **2500u Refacto AF on 22 May and check pre and post infusion level Factor VIII level and inhibitor level. For inhibitor level only one test needed.**
- **2500u Refacto AF one hour before surgery. Take pre and post samples but patient can be taken to theatre if post level on test dose on 22 May around 100iu/dl.**
- **Check factor VIII level mid surgery and top up if necessary (Discuss with Dr Ismail please)**
- **Check factor VIII level post surgery and top up if necessary (Discuss with Dr Ismail please)**
- **Will need evening dose of Refacto AF to maintain post infusion level around 100iu/dl. (please discuss the dose needed with Dr Ismail)**
- **Needs twice daily infusion of Refacto AF with pre and post levels for first 7 days post – operatively. This can be done at home if fit to be discharged.**
- **Will need daily treatment for two weeks post-operative**

Factor concentrates will be provided and prescribed by SHC. If post-procedure review is specified as "required" in the plan, please do not discharge the patient home before Haemophilia team review.

Enquiries: 9am-5pm SHC: GRO-C, SpR Bleep: GRO-C
Emergency Haematology advice is available OOH, ask for Haematology Consultant on call

GIG CYMRU NHS WALES
Bwrdd Iechyd Prifysgol
Abertawe Bro Morgannwg
University Health Board

Information for patients

Inherited Coagulation Disorders (ICDs)

Looking after your teeth and gums

Back Cover:

Helpline number/s
Haemophilia Treatment Centre (HTC) - 01792 200368
(emergency and out of hours contacts provided on answerphone)
Specialist Dental Centre (SDC) - 01792 703101 (Mon to Fri 9-5)

Document reference no:

Why are your teeth and gums important?

You may have been diagnosed with a bleeding disorder such as Haemophilia or Von Willebrands Disease. You may be a relative or carer for someone with this diagnosis.

It is important for people with bleeding or clotting (coagulation) disorders to take good care of their teeth and gums. This can reduce the chances of future problems like pain, infections or extractions. It can also reduce the need for medical treatment such as transfusion of clotting factors, or blood products.

How to I prevent dental problems?

Dental problems can be prevented by

- Practicing regular oral hygiene- brushing at least twice daily
- Reducing the frequency and amount of sugar in your diet
- Avoiding smoking
- Regular check-ups by a dentist

Attending the dentist

Your Haemophilia Treatment Centre (HTC) may have advised you about the severity of your condition. If you are not sure, just check with them (contact details on the back cover). You may have already informed your dentist about your bleeding disorder. If not, please make sure to update him/her on your next visit. Your dentist can get further help and advice from your HTC.

In most instances, if you have a bleeding or clotting disorder, it may be fine to attend your own dentist for routine care such as check ups and fillings.

However, some types of dental treatment need to be done in a specialist centre. This is because these have a higher risk of bleeding complications. These include tooth extractions, some deeper injections, gum surgery or dental implants. Your dentist should then contact your HTC for advice.

We recommend that everyone with medical conditions such as haemophilia should attend their dentist at least twice a year.

FAQs (Frequently asked questions)

Can I use my local dentist?

Many of us like the convenience of getting a full range of dental care in a convenient location. Your dentist may be able to undertake many aspects of treatment. However, this depends on the severity of your condition and the dental treatment needed. If you or your dentist would like more advice, then please contact your HTC.

Should I brush my teeth if my gums bleed?

Bleeding gums are a sign of early gum disease. Commonly, people notice this when brushing their teeth. Sometimes, people avoid brushing due to the bleeding. In fact, this makes the problem worse in the long term. If gum disease progresses, teeth can become loose and eventually be lost. We recommend a soft or medium brush with a small head. This should be used with a pea-sized amount of toothpaste at least twice a day. Electric toothbrushes can also be very effective.

What if my gums continue to bleed despite brushing regularly?

If your gums continue to bleed, then please do seek advice from your dentist. Your dentist might recommend methods for brushing and cleaning your teeth more effectively.

Your bleeding disorder may mean that your gums bleed a little more easily and for a little longer until the gums heal. Your dentist and/ or hygienist may also recommend professional cleaning. Often, patients are advised to take Tranexamic acid tablets before and after the procedure. Sometimes, Tranexamic acid is also used as a mouthwash.

Occasionally, your dentist may refer you to a Specialist Dental Centre (SDC- see contact details on back cover) for treatment.

What is Tranexamic acid?

Tranexamic acid prevents blood clots from breaking down. It is used before and after some dental procedures like cleaning (scale and polish).

What happens if I need a tooth extraction?

This must always be co-ordinated by your HTC. They will arrange this with your dentist or the SDC. They will advise about the need for factor replacement therapy based on the severity of your bleeding disorder. Tranexamic acid tablets and /or mouthwash are also normally prescribed before and after the extraction. In addition the dentist may stitch the wound and use a special surgical packing to stop any bleeding.

I cannot find a dentist locally. What should I do?

Phone your HTC on 01792 200368.
You will be given a number to contact depending on where you live. You will be assigned to one of the dedicated dental teams in the Hospital Dental Services or the Community Dental Services depending on the severity of your condition and your dental treatment needs.

My dentist seems to be unsure of treating me because of my condition. What should I do?

If you or your dentist wants further advice, then you can be referred to a SDC. Just ask your HTC about the one nearest to you.

What should I do in an emergency?

Phone your own dentist first for advice. If you are not registered or are unable to contact an emergency dentist, then phone the HTC on 01792 200368.

For more information, please visit
www.wfh.org
Oral care for people with Haemophilia or a Hereditary Bleeding tendency

Information for dentists

You must have an up to date record of the severity of your patient's bleeding disorder. Please update the medical history at every appointment.

In general, patients with mild Haemophilia and Von Willebrands Disease can safely receive most of their dental care in general dental practice. Patients with moderate or severe haemophilia (less than 5% clotting factor) will normally be treated in a specialist setting.

General Tips

The following procedures are relatively safe in patients with mild haemophilia:

Fillings- however, avoid nerve blocks and lingual infiltration
Scaling- may need to be staged and covered with Tranexamic acid if oral hygiene is poor
Root canal treatment- caution with rubber dam placement and take care to work within the anatomical apex

Handle soft tissues gently. Take care with impression trays, aspirators and x-ray films in the floor of the mouth and retromolar region.

Areas of Concern

The following procedures pose a significant risk of bleeding complications. Please refer to your SDC with an appropriate treatment plan and radiographs for the following:

Nerve blocks
Extractions
Sub-gingival debridement
Implant placement

These will need to be co-ordinated with the HTC for peri-operative management.

Local Anaesthesia

Buccal infiltration injections pose relatively little risk to patients with haemophilia. Lingual infiltrations and Inferior Dental Blocks (IDBs) however, potentially could cause deep bleeding. This might compromise the airway.

Alternative techniques to anaesthetise posterior molar teeth include
Buccal infiltration with Articaine
Intraligamentory anaethesia

If an ID Block is necessary, then patients with less than 30% factor levels will require factor replacement prior to administration.

Analgesia

NSAIDs and Aspirin have a potential to aggravate bleeding, especially in moderate to severe haemophiliacs. They must not be prescribed without the involvement of the Haemophilia Treatment Centre. They may need to normalize clotting factor levels. Paracetamol or codeine based products are more appropriate.

Concurrent illness

Patients with other conditions such as liver dysfunction must be referred to a specialist dental unit due to the increased risk of bleeding complications.

If you have any queries or concerns, then please contact
Haemophilia Treatment Centre (HTC) - 01792 200368
Or
Specialist Dental Centre (SDC) - 01792 703101

For more information, please visit
www.wfh.org
Guidelines for patients with Inherited Bleeding Disorders

References

Brewer A, Correa M.E. (2006) Guidelines for Dental Treatment of Patients with Inherited Bleeding Disorders.
World Federation of Haemophilia Monograph no. 40.

Fiske J. Ford H, Savidge G, Smith M. (2000) The expressed dental needs of patients attending a Haemophilia Reference Centre.
Journal of Disability and Oral Heath 20 (5).

Lee A, Boyle C, Savidge G, Fiske J (2005) Effectiveness in controlling haemorrhage after dental scaling in people with haemophilia by using tranexamic acid mouthwash. British Dental Journal 198:33-38.

Dougall A ,Fiske J. (2008) Special Care Dentistry-Safety.
British Dental Journal 204 (15)

GIG CYMRU NHS WALES
Bwrdd Iechyd Prifysgol
Abertawe Bro Morgannwg
University Health Board

Inherited Bleeding Disorders (IBDs)
Information for dentists

Dear Colleague,
All patients with inherited bleeding disorders should be seen for regular dental checks in general dental practice. The Haemophilia Centre can advise on how to correct haemostasis for any invasive procedure or whether the patient needs to be referred to the Specialist Dental Centre (SDC) in Morriston Hospital for a specific treatment on a case-by-case basis. Treatment might include the use of tranexamic acid or self-administering coagulation factor concentrate or desmopressin.

Most patients can safely receive most of their dental care, including invasive procedures, in general dental practice. This includes patients with moderate (1-5% clotting factor) or severe haemophilia (< 1% clotting factor) because they are usually able to self-administer treatment to correct their blood clotting system based on the advice from the Haemophilia Centre.

Prevention
Prevention is key to minimise exposure to factor replacement. There should be written documentation about the patient's oral health status and advice should be provided on preventive care at each follow-up visit,.
All children with Inherited Bleeding Disorders would be considered as high caries risk due to the complexity and morbidity of treatment if caries occurs.

General Tips
Treatment planning
Treatment planning is essential for good outcome and should involve liaison with the haemophilia centre. Carefully schedule invasive dental procedures to minimise re-exposure to factor replacement.

The following procedures are relatively safe in patients with mild haemophilia:

- Fillings; avoiding nerve blocks and lingual infiltrations and careful use of suction/aspirators.

- Supragingival scaling; but it needs to be staged and covered with tranexamic acid, if oral hygiene is poor.
- Root canal treatment, with careful rubber dam placement and working within the anatomical apex.
- Dental impressions, with careful tray placement and gentle soft tissue handling.
- Radiographs; careful positioning in the floor of the mouth and retromolar regions.

Areas of Concern
Please consult the Haemophila Centre prior to the following procedures for advice on haemostatic treatment. These procedures pose a significant risk of bleeding complications for all patients with bleeding disorder but can often be safely performed with appropriate haemostatic cover:

- Nerve blocks
- Dental extractions
- Sub-gingival root debridement
- Dental implant placement
- Minor oral surgical procedures

You can also refer to your Specialist Dental Centre with an appropriate treatment plan and radiographs.

Local Anaesthesia
Buccal infiltration, with aspirating syringes pose relatively little risk to patients with haemophilia. Lingual infiltrations and inferior dental (ID) blocks can potentially cause deep bleeding, which may compromise the airway unless the patient has had appropriate haemostatic cover.

Alternative techniques to anaesthetise posterior molar teeth include buccal infiltration with Articaine and intra-ligamentary anaesthesia. If an ID Block is necessary, then patients **with less than 30% factor levels** will require factor replacement, before administering the injection.

Analgesia
NSAIDs and Aspirin can aggravate bleeding. NSAIDs may be used on a case by case basis but **do not** prescribe without receiving advice from the Haemophilia Centre.
Paracetamol or codeine-based products are more appropriate to use.

Concurrent illness
Patients with other conditions such as liver dysfunction must be referred to a specialist dental centre due to the increased risk of bleeding complications.

If you have any queries or concerns, then please contact

Helpline number/s
Haemophilia Treatment Centre
01792 200368
(Emergency and out of hours contacts are on answer phone)

Specialist Dental Centre (SDC)
Dept of Restorative Dentistry, Morriston Hospital- 01792 703101(Mon- Fri 9 to 5)
Dept of Oral & Maxillofacial Surgery, Morriston Hospital

References

2. Anderson et al (2013) Guidance on the dental management of patients with haemophilia and congenital bleeding disorders. British Dental Journal 215:497-504.

3. Brewer A, Correa M.E. (2006) Guidelines for Dental Treatment of Patients with Inherited Bleeding Disorders. World Federation of Haemophilia Monograph no. 40.

GIG CYMRU NHS WALES
Bwrdd Iechyd Prifysgol
Abertawe Bro Morgannwg
University Health Board

Dental care for patients with Inherited Coagulation Defects (ICDs)

A quick guide for the dental team

by

Akhila Muthukrishnan

BDS MPhil FDSRCS FFDRCSI

Specialist in Special Care Dentistry

Index

Modern dental teams care for patients with many medical conditions. This workbook is designed to help the clinical dental team provide dental care for patients with **Inherited Clotting Disorders** or **ICD**s. Your role in the team may be that of a dentist, a hygienist, a therapist or a dental nurse.

Comments from patients and members of the dental team are highlighted in yellow boxes. Additional facts are presented in blue boxes by Dr Smiles. Self-assessment exercises are included at the beginning and end of the workbook. This book is also available in digital format. Finally, your comments are invaluable in designing this book. Please use the evaluation form on the last page to let us know your comments.

" I must have been 9 years old… I remember bleeding for days after my tooth was taken out…I thought I was going to die…no one knew what to do…the dentist looked so worried, I thought he would have a heart attack!"

JW, 57 yr old patient with haemophilia

- Coagulation disorders involve impairment of the Coagulation cascade (Appendix 1).
- Von Willebrand's disease is the commonest ICDs. It affects approximately 1% of the population
- Haemophilia A is the next most common, but only affects 5 in 100,000 of the population

Objectives

By the end of using this workbook, you should be familiar with the following

- The most common ICDs
- The dental aspects of ICDs
- Treatment planning for ICDs

This should help you to understand

- The importance of teamwork
- Risk management for patients with ICDs

Ultimately, this workbook is meant to supplement the 'hands-on' knowledge gained from working with experienced clinicians and patients.

Teams in the field of special care dentistry can use this to supplement their clinical experience and seek guidance from their local haematology teams for specific conditions. Even thought this workbook is designed for use in the UK, the principles apply universally.

Special Care Dentistry is the field of dentistry that promotes care for patients with physical, medical, emotional and learning disabilities

Dentistry in the UK is provided in the

- General Dental Services (GDS)
- Community Dental Services (CDS)
- Hospital Dental Services (HDS)

Initial self- assessment exercise

Read the case based scenario below and spend a few minutes to reflect on how you would manage this situation:

You are a newly appointed dentist in a 3-surgery practice. The receptionist interrupts your morning surgery to inform you that a 21 year old Haemophiliac has phoned asking to be seen urgently. He has had severe toothache for the last two days. Your nurse recognises his name and says that she knows him well. He normally sees the senior dentist in the practice and they have undertaken fillings without any problems. The senior partner is on holiday in Barbados!

How would you manage this scenario?

Write a brief summary

If you were the nurse involved, how would you deal with it?

Self- assessment questions (SAQs)

Choose the correct response

1. Haemophilia is passed on to males

   True/ False

2. von Willebrand's disease is rare in females

   True/ False

3. DDAVP is used as a mouthwash to treat bleeding

   True/ False

4. Tranexamic acid is not licensed for use by dentists

   True/ False

5. Patients with Clotting disorders must be treated in hospital

   True/ False

Responses to case based scenario on page 13

Responses to SAQs on page 14

Some common ICDs

Haemophilias

Haemophilia A is an X-linked disorder. It is related to deficiency in clotting factor VIII. It affects males. Dental extractions have led to prolonged bleeding. They have been fatal in the past.

The extent of bleeding in Haemophilia is related to the severity of the disease and the degree of trauma. Patients with mild to moderate Haemophilia can be normally treated outside the hospital environment in the Community Dental Service (CDS) or the General Dental Services (GDS). However, this also depends on the procedure being undertaken. A Haematologist must always be consulted before extractions and this may need to be done in the Hospital Dental Services (HDS). The diagnostic laboratory findings in Haemophilia are summarised in Appendix 2.

"It is really hard to get to the hospital as it is so far away. I can never get an appointment at the hospital dentist as their waiting lists are so long. I now see my local dentist, who phones my Haematologist if he needs advice. That is much easier. The only time I went to the hospital was when I had to have a tooth out."
JB, Haemophiliac

Severity of Haemophilia

	Factor VIII %
Severe	<2
Moderate	2-5
Mild	>5

von Willebrand's disease (vWD)

This is due to an inherited deficiency of von Willebrand's factor (vWF). Clinically significant vWD affects 1% of the population. It affects 6% of women with menorrhagia. It is at least twice as frequent as Haemophilia. It affects females as well as males. It is inherited usually as an autosomal dominant trait.

The nature of bleeding from vWD is similar to platelet dysfunction. The severity varies from patient to patient and from time to time. The diagnostic laboratory findings in vWD are summarised in Appendix 3.

Types of von Willebrand's disease

Type	% vWD	Defect in vWF	Fr VIIIc
1	80	Partial lack	Low
2A	15	Partial lack	Low
2B	Rare	Partial lack	Low
2M	Rare	Partial lack	Low
2N	Rare	Partial lack	Low
3	5	Complete lack	Low

Dental aspects of Haemophilias and vWD

Potential difficulties

- NSAIDs are contra-indicated- therefore pain management may be difficult
- Concurrent Hepatitis or HIV as a result of blood transfusion
- Liver disease may complicate bleeding further
- Factor VIII inhibitors
- Patients may be anxious due to frequent hospital visits in childhood
- Some patients may avoid dentists as a result of such anxiety
- Severe, even fatal bleeding from surgical procedures

General principles

- Haemophiliacs form a priority group to minimise the need for dental operative intervention
- Education of patients or parents, and preventive dentistry should be started as early as possible in the young child.
- Team working with the Haematology team is vital for risk management
- Always check and maintain contemporaneous patient records
- Seek advice from a senior colleague if necessary

"I am very lucky to work in an area where the Haematology team is so approachable. It makes all the difference to our patients. I am OK with most patients, but may need advice for the odd one. My nurse knows the Haematology nurse well now. "
KC, dentist

Caution 

- Nerve blocks should be avoided due to the risk of bleeding into spaces. Lingual infiltration should also be avoided for the same reason. A Factor VIII level of at least 30% is required if regional nerve blocks are unavoidable.
- Caution with saliva ejectors – use one with a rounded end to prevent accidental bleeding.
- Cotton rolls should be wetted before removal to prevent mucosal tears

Conservative Dentistry

Fillings can be placed without any problems in all patients with ICDs. The difficulty lies in obtaining good local anaesthesia. Caution should be exercised with matrix bands.

Orthodontics

Braces are not just to make people look pretty. They also help to make teeth even and therefore easier to clean. Team working between the Oral surgeon, Orthodontist, General Dental Practitioner and the Haematologist is essential in late childhood to co-ordinate treatment. Care must be taken to avoid sharp edges on braces.

"I was panicking when I accidentally cut the gum on this lady with vWD when I was placing a filling. My tutor showed me how to use a local haemostatic agent- Ferrous sulphate- and all was well. I will know what to do next time"
CE, dental student

Endodontics

Root canal treatment can be undertaken to avoid extractions. Intra-pulpal anaesthesia can be used to supplement infiltration and intra-

ligamental anaesthesia. Rubber dam clamps should be placed with caution. The use of apex locators is recommended to prevent over-instrumentation and extrusion of filling material.

Periodontics

Hygienists and therapists may often be called on to provide scaling for patients with ICDs. The following is advised

- Scaling can be undertaken in all but the most severe haemophiliacs outside the hospital environment
- Tranexamic acid mouthwash and tablets must be used before and after the procedure to prevent clot breakdown
- In severe haemophiliacs, the haematologist should be consulted and appropriate treatment provided before scaling.

"Tranexamic acid is a godsent! I ask my patients to rinse with a 5% solution for 2 minutes before scaling and make sure they have the tablets for a week. I do always check with the dentist if this is OK, as I know that some of the patients with bleeding disorders can be very severe. I know one who needed transfusion of factor VIII before even the simplest scaling."

LD, dental hygienist

Tranexamic acid is an antifibrinolytic and prevents clot breakdown. It can be prescribed by dentists, though the mouthwash is unlicensed.

Extractions , dento-alveolar surgery, periodontal surgery

These should be carefully planned to be comprehensive to avoid repeat visits. Patient should be referred to the Hospital Dental Services.

A Factor VIII level of 50-75% is needed for these procedures. Other measurers should include

- Tranexamic acid tablets but can also consider mouth wash
- Consider prophylactic antibiotics
- Minimal trauma to bone and soft tissues
- Primary closure with atraumatic suturing and haemostatic agents such as Surgicel™
- Careful post-operative care to prevent late post-operative bleeding

Haemophilias		
Factor VIII level required	Pre-operatively give	Post-operative
	Factor VIII Tranexamic acid 1g oral TDS starting the day prior to dental intervention and continue for 3-7 days after depending on intervention	Local haemoststic measures

Von Willebrand's disease

Type of vWD	Desmopressin	Factor VIII
1	IV, or SC	-
2A	IV or SC	=/-
3	Contra-indicated	+

Response to case based scenario

Anyone who has worked in practice knows the pressures of interruptions. The key features of this scenario are

- This patients is in severe pain
- Normal painkillers such as NSAIDs cannot be taken
- There may be difficulties with obtaining local anaesthesia

Your management should include the following

- Check his previous records to verify the severity of Haemophilia
- Check his previous dental treatment and complications, if any
- Work as a team- the experienced nurse can be very helpful in calming the patient down
- Choose the safest mode of local anaesthesia
- Provide emergency treatment to alleviate pain, schedule at the end of a session
- Phone the local haematologist, if in doubt
- The senior dentist can take over on his return from Barbados!

How did you do?

If you considered the key features, then you are on your way! As these cases are relatively rare in General Practice you may want to read further (see page 18-20). You may wish to discuss this scenario with a senior colleague and ask how they would manage it

Self assessment responses

1. Haemophilia A / B manifest clinically in males

   True

   This is a sex-linked inheritance pattern. Females can be carriers, but rarely manifest the disease.

2. von Willebrand's disease is rare in females

   False

   vWD is twice as common as haemophilia. A common route of diagnosis is when women present with menorrhagia.

3. DDAVP is used as a mouthwash to treat bleeding

   False

   DDAVP is used as IV, SC or nasal spray. It helps to release Factor VIIIc, vWF and tPA from the endothelium

4. Tranexamic acid is not licensed for use by dentists

   False

   Tranexamic acid tablets can be prescribed for oral use by dentists. The mouthwash form can only be obtained through hospital pharmacies, as it is an unlicensed preparation.

5. Patients with Clotting disorders must be treated in hospital

   False

   Most patients with clotting disorders can obtain most of their dental treatment outside the hospital environment. Patients with severe ICDs, those requiring nerve blocks and those requiring extractions or dento-alveolar surgery need consideration for hospital treatment.

How did you do?

If you scored 2 or below on the self assessment questions, you may find it useful to review the reading material. Ensure you have grasped the principles before attempting the next self- assessment MCQs. If you scored 5/5 excellent, now you need to extend your knowledge base by looking at the suggested reading and guidelines in the websites on page 18-20. Then try the next challenge!

"I enjoy treating patients with complex medical problems- it gives you such a feeling of satisfaction to know that you have looked after them well. And the patients are so grateful, because they have so much else going on with their lives. "

TG, dental nurse

MCQs

1. Which of the following is safe for patients with ICDs
    a. Paracetamol
    b. Codeine
    c. Aspirin
    d. Brufen
2. Local anaesthesia for patients with ICDs can include
    a. labial infiltration
    b. lingual infiltraction
    c. nerve blocks
    d. intra-papillary injections
3. Haemophiliacs with inhibitors can be treated with
    a. DDAVP
    b. FEIBA
    c. Factor VIIIc
    d. Tranexamic acid
4. In assessing VWD , the following are needed
    a. VIIIc assay
    b. Ristocetin-cofactor
    c. VII
    d. Factor VIII; VWF antigen
5. Local haemostatic measures include
    a. suturing the socket
    b. ferrous sulphate
    c. surgicel
    d. all of the above

Answers

1. a,b 2. a,d 3. a,b,d 4. a, b &d 5. d

Summary

- **Patients with ICDs are a high priority group**
- **Most patients with ICDs can undergo a wide range of dental treatment safely outside a hospital environment**
- **Patients with severe haemophilia may need hospital based care**
- **Nerve blocks should be avoided. If they are unavoidable, then a Factor VIII level of at least 30% is needed.**
- **Extractions, dento-alveolar surgery and periodontal surgery need careful planning. Factor levels need to be checked and appropriate cover provided**
- **Team-working with the Haematology team is essential**
- **Prevention is vital to minimise the need for treatment**

Further reading

1. Sculley C, Cawson RA. (2005) *Medical problems in Dentistry*. Elsevier Churchill Livingstone, London, UK.
2. Sculley C, Dios PD, Kumar N. (2007). *Special Care in Dentistry.* Churchill Livingstone, London, UK.

Useful Websites

1. British society of Haematology- www.b-s-h.org.uk
    - Prophylaxis in children and adults with Haemophilia
    - Guidelines for the management of patients on oral anticoagulants requiring dental surgery

2. UK Haemophilia society- www.haemophilia.org.uk
    - Patient information
    - Useful contacts

Appendix 1

The Coagulation cascade is in the secondary stage of haemostasis (blood clotting). The aim of secondary haemostasis is the formation of fibrin. The primary stage involves vascular and platelet activity. The coagulation cascade involves Coagulation factors acting as enzymes, which require activation and co-factors. Calcium and platelets are also required along with a binding surface.

Defects in primary haemostasis cause more serious bleeding than defects of primary haemostasis. They include bleeding into cavities (cranium, chest, joints) and sub-cutaneous haematomas.

Blood coagulation factors	
Factor	**Name**
I	Fibrinogen
II	Prothrombin
VII	
VIII	Antihaemophilic Factor
IX	Christmas Factor
X	Stuart-Prower Factor
XI	Plasma Thromboplastin Antecedent
XII	Hageman Factor
XIII	Fibrin Stabilising Factor
Fitzgerald Factor	High molecular weight kininogen
Fletcher Factor	Prekallikrein

Appendix 2

Laboratory findings in haemophilia

- Prolonged Activated Partial Thromboplastin Time (APTT)
- Normal Prothrombin Time (PT)
- Normal Bleeding Time
- Low Factor VIII c but normal Factor VIIIR:AG (von Willebrand factor) and R:RCo (Ristocetin co-factor)

Factor VIII assay is required as even the APTT may be normal in mild haemophilia

Appendix 3

Laboratory findings in von Willebrand's disease

- Prolonged bleeding time
- Usually a prolonged APTT
- Low levels of von Willebrand's factor (factor VIIIR:Ag)
- Low Factor VIIIC
- Low VIIIR: RCo (Ristocetin co-factor)

Unlike haemophilia, the bleeding time is prolonged. The best assay is ristocetin co-factor assay.

Evaluation form

Please complete the evaluation form and post/ e-mail to the address below.

1. As a result of using this workbook, I feel more confidant in managing patients with ICDs
    a. Completely agree
    b. Partly agree
    c. Disagree

2. Please comment on what was the most useful part of this workbook.

.....

3. Please comment on what was the least useful part of the workbook.

.....

4. Overall, I feel I have gained more knowledge about the subject matter
    a. Completely agree
    b. Partly agree
    c. Disagree

Any other comments

.....

.....

For an e-version of this workbook, please e-mail Akhila.muthukrishnan@ GRO-C