Nursing Management of Haemophilia

and von Willebrand Disorder

Australian Haemophilia Nurses Group

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Haemophilia

For the purpose of this document, the term "haemophilia" refers to FVIII / FIX deficiency and other coagulation disorders.

Haemophilia is a blood clotting disorder in which one of the essential clotting factors is deficient. Haemophilia A or classical haemophilia, is due to the deficiency of factor VIII. Haemophilia B (or Christmas disease) is due to the deficiency of factor IX. As a result of a deficiency of these clotting factors, formation of a blood clot is delayed and prolonged bleeding may occur after minor injury or sometimes spontaneously. Haemophilia is a problem of secondary haemostasis and is hereditary. Haemophilia A occurs in approximately 1 in 8000 male births. Haemophilia B is less common and occurs in approximately 1 in 30,000 male births. Haemophilia is an X linked disorder and affects males almost exclusively, although females can be affected either by carrying the haemophilia gene (sometimes referred to as symptomatic carriers) or by having both x chromosomes affected by haemophilia mutations.

Haemophilia A is managed by either replacing the deficient factor VIII with a FVIII concentrate or in mild cases using a drug called Desmopressin Acetate (DDAVP) to increase the circulating factor VIII. Patients should have a proven response to DDAVP before it is considered for treatment.
Haemophilia B is managed by replacing the deficient factor IX, with a FIX concentrate.

For further instructions see below – Administration of replacement therapy with Factor concentrates.

Von Willebrand Disorder

Von Willebrand Disorder is the most common hereditary bleeding disorder and may occur in up to 1% of the population. It is mostly an autosomal dominant disorder, but can also be inherited as an autosomal recessive disorder. Patients with von Willebrand Disorder either have diminished production of von Willebrand factor or produce von Willebrand molecules that do not function normally – hence their platelets do not adhere properly when blood vessels are injured and it takes longer for bleeding to stop. In some patients, Factor VIII is also reduced, and blood clotting is impaired. Von Willebrand Disorder is a problem of primary haemostasis.

Von Willebrand Disorder can in some cases be managed with DDAVP as described above. DDAVP, as well as increasing the circulating factor VIII, also increases the circulating von Willebrand factor. Depending on severity of the von Willebrand Disorder, plasma concentrates which contain both von Willebrand factor and factor VIII may be required as the replacement therapy.

For further instructions see below – Administration of replacement therapy with Factor concentrates.

Administration of replacement therapy with factor concentrates

- Australian states and territories may differ regarding where replacement therapies may be obtained from. These include the hospital blood bank, haemophilia centre, hospital pharmacy or Australian Red Cross Blood Service.

- Replacement therapy products should ideally be stored in the refrigerator until required, however can be stored at room temperature below 25° Celsius. Storage outside the refrigerator will reduce the shelf life. Please read the individual package insert. It is important that the product is not frozen.

- When checking the product, ensure the correct product is administered to the patient. The patient should be treated using the same brand as that prescribed by the patient's haemophilia haematologist, unless in a life threatening situation. If in doubt call your closest Haemophilia Centre. Record batch numbers by peeling batch labels off the bottles. Whole bottles of factor should be administered and doses should be rounded up.

- Replacement therapy is given as a slow push intravenously as per package insert. **It is important to follow package inserts re the reconstitution of these products.**

- When reconstituting products, use the diluent provided. Do not dilute further. DO NOT SHAKE

- Any reactions should be reported to the local Haemophilia Centre

- The half life of factor VIII is 8 to 12 hours. Therefore twice daily dosing may be required. The half life of factor IX is 18 to 24 hours. Therefore daily dosing may be required. If there are doubts as to how much treatment a patient may require, please contact your local Haemophilia Centre

DDAVP

DDAVP is given either as an intravenous infusion over 20 – 30 minutes dependent on local regimens, or subcutaneously. Patients should be fluid restricted, when having DDAVP to avoid hyponatraemia side effect. Tachyphylaxis may occur after 3 doses.
Note: May not be suitable for with patients with cardio-vascular disease or who are pregnant

Complicating conditions

Unfortunately, people with haemophilia often face additional challenges associated with their bleeding disorder. In many instances, it is these additional complications that will bring the patient to your hospital. The complications are either as a result of past blood product infusion or as a result of repetitive bleeds.

The most common complications encountered are

- Hepatitis B & C / liver disease
- HIV & AIDS
- Joint arthropathy causing loss of mobility and/or pain
- Antibody formation to factor products (known as 'inhibitors')

Care in the Emergency Department

Normal first aid measures should be used.
Emergency treatment is needed when a patient suffers trauma/injury, a bleeding episode or displays signs/symptoms indicating a possible bleeding event. Patients with severe bleeding disorders can present with spontaneous bleeding events with no precipitating cause.

Key Points

- Check if the patient has an Emergency Treatment Card. All patients are supplied a card from their Haemophilia Treatment Centre (HTC), displaying their details, type and severity of the bleeding disorder, and recommended treatment.

- The patient should be seen urgently and factor treatment administered before further investigations are undertaken (such as x-rays, CT scan, scoping). Delays in receiving treatment can increase morbidity and the risk of mortality

- Patients on home therapy programs may have treated themselves with factor before presenting to ED, if so check the factor dose to ensure it is therapeutic

- Sometimes patients will bring their own factor supply from home with them (this is more likely if they have severe haemophilia). If so, this should be used. If not, ask them which factor brand they use as there is a range of products.

- Calculate the required dose as sometimes the patient will suggest doses that may be sub therapeutic for the type of injury.

- In the absence of a documented baseline factor level dosing is calculated as if the factor level is 0%
- As baseline factor levels remain unchanged, factor levels do not need to be checked before factor treatment is initiated
- R.IC.E. – Rest, Ice, Compression and Elevation are simple measures which can be beneficial
- Avoid intramuscular (IM) injections and NSAID's.
- Lumbar punctures and other such invasive needles should only be performed after an appropriate dose of factor has been administered
- If significant trauma has occurred and the patient can demonstrate his haemophilia diagnosis, do not wait for obvious signs of bleeding
- In most circumstances patients will require more than one dose to treat a bleed
- If in doubt, treat with appropriate factor immediately.
- In most cases, the haemophilia patient is well aware of the correct treatment required for his disorder and his opinions and comments should be treated with respect.
- Patients may be very protective of their veins and are often expert at the placement of intravenous needles as they self access them during home therapy use of factor VIII or factor IX.
- A haemophilia patient may become assertive if treatment is delayed. This is because he has been educated by his haematologist to seek a factor VIII or IX infusion as an urgent measure.

Life Threatening or limb threatening Bleeds	Nursing Actions Required	Comments
Central Nervous System Compartment Syndrome Gastrointestinal Neck/Nose/Throat Severe Injury Hip or iliopsoas Advanced joint/muscle	• Assessment (appropriate for site) • Vital Signs • Administer factor ASAP • analgesia	**Immediate factor replacement is essential. Raise the factor level to 80 - 100%** Contact the HTC staff or haematologist for ongoing treatment advice
Non Life-Threatening Bleeds		
Joint Muscles/soft tissue	• Assessment: site, pain, swelling, ROM • Vital Signs • R.I.C.E.	For patients with mild haemophilia A or type 1 vWD, Desmopressin (DDAVP) may be considered instead of factor concentrates. Raise the factor level to 50 - 60%
Haematuria	• Assessment: pain, degree of bleeding • Fluids +++ • Bed rest	Factor replacement and anti-fibrinolytic agents (eg Tranexamic acid) are **contraindicated** until consultation with HTC
Mucosal Bleeding Mouth/gums Mild –Moderate Epistaxis	• Assessment: degree of bleeding • Local measures	Anti-fibrinolytics (topical or oral) are first line of treatment & should be considered alongside DDAVP/factor concentrates

Laboratory Testing

Diagnosis	Laboratory Tests	Comments
Haemophilia A	APTT Factor VIII assay	The APTT is prolonged. In symptomatic carriers the factor VIII level usually rises during pregnancy
Haemophilia B	APTT Factor IX assay	The APTT is prolonged
Von Willebrand Disorder	Factor VIII complex • Factor VIII assay • VWF-Ag • VWF-RiCof • VWF-CBA Platelet function analysis	The factor VIII complex gives a complete picture on the amount and functionality of von Willebrand factor. During pregnancy the factor VIII and VWF may rise (except in type 2&3 VWD). * Note that the APTT is not used in VWD as it is usually normal (except in severe patients) * Call lab in advance to advise if a platelet function test is required. Check for hospital specific details re collection & delivery of samples
Results related to rare bleeding disorders	Prolonged APTT	FXI or FXII deficiency
	Prolonged Prothrombin Time (PT)	FVII deficiency
	Prolonged APTT and PT	Common pathway deficiency i.e. FII, FV or FX deficiency

Please note that only main laboratories perform factor testing (Factor VIII, IX, Factor VIII complex and Platelet Function Analysis). Therefore these samples should only be taken if they can reach the main laboratory ideally within 2 hours.

Surgical and Inpatient Care

Prior to any invasive procedure patients with a known bleeding disorder should be reviewed by a haematologist or by the team at a Haemophilia Treatment Centre. It is the responsibility of the haematologist / HTC Team to prescribe the necessary bleeding disorder treatment and possible blood tests required. Treatment may be required for up to 10 days.

DDAVP (Desmopressin) is usually available from Pharmacy. Factor concentrates are located in either the Pharmacy or Blood Bank (Transfusion Medicine Unit), depending on your hospital's preference.

Treatment is generally given within1 hour pre operatively unless stated otherwise.

Patients should be monitored for bleeding post procedure. Remember that the treatment administered corrects the patients bleeding disorder to within the "normal range". Therefore some bleeding may be considered normal. Excessive bleeding or changes in patients' vital signs warrant urgent review.

TEDS and foot/calf pumps are appropriate post operative measures in the prevention of DVT associated with surgery. The decision to use anti-coagulant DVT prophylaxis (e.g. heparin) should be made in consultation with the haematologist.

Patients may require follow-up treatment in the post-operative period. Necessary treatment and blood tests required should be ordered either by or in consultation with a haematologist.

If you have any questions about a patient having surgery please contact your relevant Haemophilia Treatment Centre.

Discharge Planning

The Haemophilia Treatment Team should be involved with the patient's discharge planning. Relevant to considerations are:

- Need for ongoing coagulation factor replacement (supply and administration)
- Wound care / CVAD care
- Further blood test monitoring
- Follow up reviews

Other Nursing Considerations

General Care	Key Points
No intramuscular injections	Muscles are very vascular and the risk of bleeding and abscess formation is great
No Salicylates / aspirin or products containing aspirin eg NSAIDS	Causes platelet malfunction and side effects such as gut bleeds
No invasive procedures without first replacing the missing factor	Includes: removal of sutures, biopsy, lumbar puncture, dental and all surgical procedures including cautery
Apply pressure x 3-5 minutes post venipuncture, de-accessing of portacaths and removal of intravenous lines	Many patients treat themselves intravenously using butterfly needles. It is essential that good vein health is maintained so they can access their own veins
Treatment & pain relief should be given promptly and before investigations	Signs of bleeding may not initially be apparent, but can result in pressure on nerve endings and in joint spaces. The sooner treatment commences and bleeding is arrested, less blood volume will be exerting pressure
Patient (or parent) is a good resource	Check for Emergency card - which often has treatment instructions

Australian Bleeding Disorders Registry

All patients receiving either recombinant factors or plasma factor concentrates should be registered on the Australian Bleeding Disorders registry (ABDR). This database collects clinical information and records factor replacement therapy usage through out Australia. If patients are not entered on this database they may potentially not be funded for replacement factor products by the National Blood Authority. Please contact your local HTC for further information.

Links / further references

Srivastava, A et al. Guidelines for the management of haemophilia.
http://www1.wfh.org/publications/files/pdf-1472.pdf