NURSING GUIDELINES FOR THE TREATMENT OF HEMOPHILIA & OTHER INHERITED BLEEDING DISORDERS

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INTRODUCTION

This information packet has been prepared to assist nurses who are unfamiliar with the care of hemophilia and other bleeding disorders. It contains general information that cannot be construed as specific advice to an individual nurse or individual patient. Conditions such as Hemophilia A, Hemophilia B and von Willebrand Disease will be defined and general guidelines for nursing care will be outlined. Our goal is to keep the document short and concise. Further and more complete information can be obtained by contacting the Bleeding Disorders Program at 519-685-8500 extension 53582 or through E-mail: sworhp@lhsc.on.ca.

Early detection and treatment of a bleeding episode can be vital for the survival of the patient.

Comprehensive Hemophilia Treatment Centres (HTC) follow most people with inherited bleeding disorders. Please contact the patient's HTC for complete medical and nursing history to ensure that s/he will receive appropriate follow-up. For a complete Canadian HTC directory please visit
Treatment Centres - Canadian Hemophilia Society
For an international directory please visit
Passport - World Federation of Hemophilia

There are many clotting proteins involved in hemostasis. Any one of these could be missing or malfunctioning leading to prolonged or excessive bleeding. The most common inherited bleeding disorders are Hemophilia and von Willebrand disease.

HEMOPHILIA

Hemophilia is a hereditary bleeding disorder characterized by decreased function or absence of factor VIII (classic hemophilia or hemophilia A) or factor IX (Christmas disease or hemophilia B). 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 spontaneously in patients with severe disease (<1%). Contrary to popular belief, a person with hemophilia does not bleed faster he bleeds for a longer period of time.

VON WILLEBRAND DISEASE

Von Willebrand disease (VWD) is the most common hereditary bleeding disorder and occurs in up to 1 percent of the population. Patients with VWD have diminished production of von Willebrand factor or produce a molecule that does not function normally. Platelets do not adhere properly when blood vessels are injured therefore it takes longer for bleeding to stop. In some patients, factor VIII is also reduced, and blood clotting is impaired.

MAIN CLINICAL AND LABORATORY FINDINGS IN HEMOPHILIA A, HEMOPHILIA B AND VON WILLEBRAND DISEASE

	Hemophilia A	**Hemophilia B**	**Von Willebrand Disease**
Inheritance	sex-linked recessive	sex linked recessive	autosomal dominant
Main sites of hemorrhage	muscles, joints, post-trauma or surgery	muscles, joints, post-trauma or surgery	mucous membrane, post-trauma or surgery
Platelet count	normal	normal	normal
Bleeding time	normal	normal	normal to prolonged
Prothrombin Time (PT)	normal	normal	normal
Partial thromboplastin time (PTT)	prolonged	prolonged	usually normal, prolonged in severe cases
FVIIIC	low	normal	low-normal
VWF: Antigen	normal	normal	low
VWF: Activity	normal	normal	low
FIX	normal	low	normal

IMPORTANT

GENERAL NURSING GUIDELINES

- NO IM INJECTIONS
- NO INVASIVE PROCEDURES WITHOUT FIRST REPLACING MISSING FACTOR
- APPLY PRESSURE X 3-5 MINUTES POST VENIPUNCTURE
- “FACTOR FIRST” TREAT FIRST, INVESTIGATE LATER

IMPORTANT

TREATMENT OF HEMOPHILIA

Clotting Factor Concentrate currently used by Bleeding Disorders Program
CSL Behring: Humate P (plasma derived Von Willebrand factor & factor VIII) **Bayer:** Kogenate FS (recombinant factor VIII) **Baxter:** Advate (recombinant factor VIII) **Baxter:** Factor VII (plasma derived factor VII) *NB: Special Access Product* **Pfizer:** BeneFIX (recombinant factor IX) **Novo Nordisk:** Niastase RT (recombinant factor VIIa)

Management of Bleeding Episodes using Clotting Factor Replacement Therapy

Hemorrhagic Event	Factor VIII, Hemophilia A	Factor IX, Hemophilia B	Factor VIII & IX with Inhibitors
<u>Mild Hemorrhage</u> • early joint, muscle • severe epitasis • persistent hematuria • gingival or dental bleed unresponsive to Tranexamic Acid	25 units/kg 50%	50 units/kg 50%	See Medic Alert
<u>Moderate Hemorrhage</u> • advanced joint or muscle bleed • dental surgery			
<u>Severe Hemorrhage</u> • life or limb threatening bleed • head injury • trauma • surgery • hematoma of neck, tongue or pharynx • compartment syndrome	50 units/kg 100%	100 units/kg 100%	See Medic Alert

***** Call the Bleeding Disorders Program immediately when bleed is severe or not responding.**

ADMINISTRATION GUIDELINES

Right Drug
Obtain factor concentrate from Blood Transfusion Services as per BDP Treatment Protocol (available in Power Chart under Blood Product Information tab and in binders located in ER & Blood Transfusion Services Departments).

Right Dose
Prior to reconstitution verify the lot number, expiry date & dosage issued. Dose to the nearest vial. Check dose against BDP Treatment Protocol.

Right Patient
Verify the vial with the patient or parent to ensure it contains the appropriate factor replacement for this patient.

Right Route
Use #23 or 25-gauge butterfly or existing IV site (flush existing IV with NaCl prior to administration by IV direct push). Ask patient for desired site.

Right Rate
Administer the dose by bolus infusion over a period of less than 5 minutes.

THINGS TO REMEMBER

- Patient is usually aware of bleeding before clinical signs are apparent
- ~250, 500, 1000, 1500, 2000 & 3000 unit vials of factor VIII and factor IX are available.
- Dose to the nearest vial.

WHEN IN DOUBT, INFUSE!