

Remember... FactorFirst

PROMPT INFUSION will halt bleeding, minimize long-term complications and can save life. If bleeding persists, follow the guidelines for life or limb-threatening bleeds and call the:

Hemophilia Treatment Centre

Physician: _____

Nurse: _____

Day Phone: _____

Night Phone: _____

Delay in the restoration of hemostasis to the patient with hemophilia or von Willebrand disease may be life or limb-threatening.

• PROMPT TRIAGE AND ASSESSMENT.

- Determine the severity of the bleed.
- Recognize that bleeding in the head, spine, abdomen or pelvis may initially be occult and potentially life-threatening.
- **TREAT FIRST AND INVESTIGATE LATER – “FACTOR FIRST”.**
- Avoid invasive procedures such as arterial punctures unless the patient has factor replacement.
- **NO** IM injections and **NO** ASA.
- The patient or guardian may be your most important resource, so do ask about specific treatment protocols.
- Contact the patient’s Hemophilia Treatment Centre where a hematologist is always on call.
- Provide clear discharge instructions and arrange a follow-up plan or admit to hospital if necessary.

Patient Information:

Name: _____

Date of Birth: _____

Diagnosis: _____

Severity: _____ Level: _____

Response to desmopressin (DDAVP): no yes to _____ %

Inhibitors: no yes

Other Medical Information: _____

Date of Recommendation: _____ / _____ / _____

Signature of Physician _____

Recommended Treatment:

Product and Dose/kg for Life or Limb-threatening Bleeds:

Product and Dose/kg for Moderate/Minor Bleeds:

Use Universal Precautions

GUIDELINES FOR EMERGENCY MANAGEMENT OF HEMOPHILIA AND VON WILLEBRAND DISEASE

LIFE OR LIMB-THREATENING BLEEDS

- Head (intracranial) and neck
- Chest, abdomen, pelvis, spine
- Iliopsoas muscle and hip
- Massive vaginal hemorrhage
- Extremity muscle compartments
- Fractures or dislocations
- Any deep laceration
- Any uncontrolled bleeding

MODERATE/MINOR BLEEDS

- Nose (epistaxis)
- Mouth (including gums)
- Joints (hemarthroses)
- Menorrhagia
- Abrasions and superficial lacerations

TREATMENT FOR LIFE OR LIMB-THREATENING BLEEDS

PATIENT MUST RECEIVE PRODUCT URGENTLY

Hemophilia A: (all severities)

Recombinant factor VIII concentrate 40-50 units/kg

Hemophilia B: (all severities)

Recombinant factor IX concentrate 100-120 units/kg >15 yrs
Recombinant factor IX concentrate 135-160 units/kg <15 yrs
The dosage for recombinant factor IX is substantially higher because of its lower recovery, particularly in children.

Von Willebrand Disease:

A VW factor concentrate containing factor VIII such as Humate-P 60-80 Ristocetin cofactor units/kg

It is critical to raise the factor level to 80-100% urgently for all life or limb-threatening bleeds.

TREATMENT FOR MODERATE/MINOR BLEEDS

PATIENT MUST RECEIVE PRODUCT WITHIN 30 MINUTES WHENEVER POSSIBLE

Hemophilia A: (severe/moderate)

Recombinant factor VIII concentrate 20-30 units/kg

Hemophilia A: (mild)

Desmopressin (Octostim/DDAVP) 0.3 mcg/kg (max. 20 mcg) –SC/IV

Hemophilia B: (severe/moderate/mild)

Recombinant factor IX concentrate 35-50 units/kg >15 yrs
Recombinant factor IX concentrate 50-70 units/kg <15 yrs
The dosage for recombinant factor IX is substantially higher because of its lower recovery, particularly in children.

Von Willebrand Disease:

Type 1 and Type 2A or 2B known to have used desmopressin safely and effectively – (Octostim/DDAVP) 0.3 mcg/kg (max. 20 mcg) –SC/IV

For patients not responding to desmopressin (such as Type 3 or Type 2B) use a VW factor concentrate containing factor VIII such as Humate-P 60-80 Ristocetin cofactor units/kg

For mucosal bleeds in all above add:

Tranexamic Acid (Cyklokapron) 25 mg/kg po tid 1-7 days (contraindicated if hematuria)

Dosages are patient specific – these are general guidelines only. Round doses up to the nearest vial. If the products listed are not available, please call the nearest Canadian Blood Services or Héma-Québec Centre.

FactorFirst



Canadian Hemophilia Society
Help Stop the Bleeding



Association of Hemophilia Clinic Directors of Canada

www.hemophilia.ca/emergency