GUIDELINES FOR EMERGENCY MANAGEMENT OF HEMOPHILIA AND VON WILLEBRAND DISEASE

FactorFirst

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.

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.

For more detailed guidelines on emergency hemophilia care please consult:
www.hemophilia.ca/emergency

Canadian Hemophilia Society
Help Stop the Bleeding

AHCDC Association of Hemophilia Clinic Directors of Canada

HEMOPHILIA TREATMENT CENTRE: