## MAJOR BLEEDS

- Head (intracranial), ocular and neck (throat)
- Spinal cord
- Intra-abdominal
- Iliopsoas muscle
- Massive vaginal hemorrhage
- Gastrointestinal

## MODERATE/MINOR BLEEDS

- Deep lacerations
- Nose (epistaxis)
- Oral (especially tongue)
- Joints (hemarthroses)
- Muscle compartments
- Menorrhagia

## TREATMENT FOR MAJOR BLEEDS

**Hemophilia A (all severities)**
One (1) unit per kilogram of recombinant factor VIII concentrate generally provides a rise of 2% FVIII activity level.
Standard dosing for major bleeding of recombinant FVIII concentrate is 40-60 units/kg.
If known desmopressin responder (see reverse side of card): desmopressin 0.3 mcg/kg SC/IV.

**Hemophilia B (all severities)**
One (1) unit per kilogram of recombinant factor IX concentrate generally provides a rise of 0.6-1.0% FIX activity level.
Standard dosing for major bleeding of recombinant FIX concentrate is 80-140 unit/kg.
Refer to product monograph for dosing instructions specific to factor replacement product.

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

**Von Willebrand disease**
A von Willebrand factor concentrate containing factor VIII such as **Humate-P** 60-80 Ristocetin cofactor units/kg or **Wilate** 40-60 units/kg.
Desmopressin could also be considered for some patients if an adequate response is documented.

## TREATMENT FOR MODERATE/ MINOR BLEEDS

**Hemophilia A (all severities)**
One (1) unit per kilogram of recombinant factor VIII concentrate generally provides a rise of 2% FVIII activity level.
Standard dosing for moderate/minor bleeding of recombinant FVIII concentrate is 20-40 units/kg.
If known desmopressin responder (see reverse side of card): desmopressin 0.3 mcg/kg SC/IV.

**Hemophilia B (all severities)**
One (1) unit per kilogram of recombinant factor IX concentrate generally provides a rise of 0.6-1.0% FIX activity level.
Standard dosing for moderate/minor bleeding of recombinant FIX concentrate is 40-60 units/kg.
Refer to product monograph for dosing instructions specific to factor replacement product.

**Von Willebrand disease**
Desmopressin SC/IV. Standard dosing is 0.3 micrograms per kg.
A von Willebrand factor concentrate containing factor VIII such as **Humate-P** 40-60 Ristocetin cofactor units/kg or **Wilate** 20-40 units/kg.
Please note: Desmopressin is NOT a suitable medication for VWD Type 2B or Type 3 patients.

**Dosages are patient specific – these are general guidelines only. Refer to product monograph for dosing instructions. Round doses up to the nearest vial. If the products listed are not available, please call the treatment centre team for advice around suitable alternatives.**

# GUIDELINES FOR EMERGENCY MANAGEMENT OF HEMOPHILIA AND VON WILLEBRAND DISEASE

# FactorFirst

Canadian Hemophilia Society
Help Stop the Bleeding

AHCDC Association of Hemophilia Clinic Directors of Canada

**www.hemophilia.ca/emergency**

## Remember …

# FactorFirst

**TREATMENT should be given in a timely manner to stop bleeding, improve outcomes and speed up recovery. Contact the care team below for treatment recommendations and support in the management of this patient.**

### Bleeding disorder treatment centre

Hospital:

Physician(s):

Nurse:

Phone:

After hours contact:

E-mail:

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### PROMPT TRIAGE AND ASSESSMENT

- Determine the location and severity of the bleed.
- Strongly consider factor replacement **PRIOR** to diagnostic procedures or consultation/detailed examination. Early treatment can mitigate further bleeding concerns or complications.
- Recognize that bleeding in the intracranial and intra-abdominal bleeding may be occult and inciting injury may have happened in days prior to presentation.
- With invasive procedures (i.e. arterial punctures, intubation) clotting factors should be normalized with replacement therapy.
- Please note aPTT will likely be shortened if patient is on emicizumab (Hemlibra) and is expected. If coagulation tests are needed, please consult hematology for advice.
- Contact the patient's bleeding disorder treatment centre where a hematologist is always on call. Patients may be very knowledgeable about their bleeding disorder and be able to provide information.
- Communicate ER visit, hospitalization to the patient's bleeding disorder care team.
- Mild disorders can develop serious bleeding in certain circumstances.

### Patient information:

Name:

Date of birth:

Diagnosis:

Severity: Level:

Response to desmopressin (DDAVP): ☐ no ☐ yes to ____%

Inhibitors: ☐ no ☐ yes

Patient on emicizumab (Hemlibra): ☐ 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: