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 bleeding disorder 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:**

### Recommended Treatment:

Product and dose/kg for life or limb-threatening bleeds:

Product and dose/kg for moderate/minor bleeds:

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**Use Universal Precautions**
**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

**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 VWF factor concentrate containing factor VIII such as Humate-P 60-80, Ristocetin cofactor units/kg or Wilate 40-60 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 safety 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 VWF factor concentrate containing factor VIII such as Humate-P 40-60 Ristocetin cofactor units/kg or Wilate 20-40 units/kg.

*For mucosal bleeds in all above add:*
Tranexamic Acid (Cyklokapron) 25 mg/kg po tid 1-7 days (contraindicated if hemorrhagia)

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 Hema-Quebec Centre.