TRAUMA CHEZ UN HÉMOPHILIE

Mécanisme du trauma

Négligeable

Évaluation clinique

Traitement selon l'avis hématologue de garde

Administrer facteur avant vaccin

Non négligeable ou suspicion clinique saignement**

LA PRIORITÉ EST D'ADMINISTRER LE FACTEUR DÉFICITAIRE dès que possible (<1h de l'arrivée)

80% des hémophiles sont hémophiles A sans inhibiteurs (=besoin de rFVIII)

Assigner personne responsable pour:

1. Avis hématologue **de l'hémostase** de garde (514 345-2360)
2. Déterminer **(sur carte « Facteur d'abord » ou Quanum):**
   - Type hémophilie A / B ?
   - Niveau de facteur?
   - Présence d'inhibiteur (faible <5BU / élevé >5BU) ?
   - Emicizumab (Hemlibra)?
3. Voir **tableau pour dose** (verso)
4. Utiliser la **médication de la famille** si disponible

Activation trauma selon la situation
Favoriser **2 accès IV** (éviter membre traumatisé)
Bilan trauma habituel (pas nécessaire de doser facteur initialement)

Etat clinique stable

ADMINISTRER LE FACTEUR DEFICITAIRE
+
Acide tranexamique 10mg/kg IV

Etat clinique instable

- **ADMINISTRER LE FACTEUR DEFICITAIRE**
- Activation trauma **niveau 1**
- Prise en **charge classique choc hémorragique (PHM)**
- **Acide tranexamique 30mg/kg IV (max 2000mg)**

Pour choc réfractaire

- **Perfusion continue de facteur** après dose initiale à discuter avec hématologue

- Investigations des sources de saignement à discuter avec hématologue

**Suspicion de saignement à n'importe quel endroit (cérébral, cervical, ORL, thoracique, abdominal, articulation, muscle) ou blessure importante (tête, cou, ORL, yeux, thorax, abdomen, membres, fracture/luxation, plaie ouverte) ou nécessitant procédure ou chirurgie invasive.

Groupe de travail en traumatologie & Dresse. Paola Rouge Elton, Mélanie Labrosse, Natalie Mathews CHU Sainte-Justine— Oct 2025

Doses et facteurs à administrer

	Hémophilie A	Hémophilie B
Taux inhibiteur ABSENT ou FAIBLE (<5BU)		
Saignement mineur/ modéré ou chirurgie mineure *(articulations, muscles superficiels, lacérations profondes, épistaxis, reins)*	**Facteur rVIII (rFVIII)** 25 IU/kg IV (administration sur 2 min)	**Facteur rIX (rFIX)** 50 IU/kg IV (administration 10ml/min)
Saignement sévère ou chirurgie majeure *(cérébral, intra-abdominal, gastro-intestinal, gorge, cou, muscles ilio-psoas ou profonds avec lésion neurovasculaire ou perte de sang importante)*	**Facteur rVIII (rFVIII)** 50 IU/kg IV (administration sur 2 min)	**Facteur rIX (rFIX)** 100 IU/kg IV (administration 10ml/min)
Thérapie adjuvante	**Desmopressine** 0.3mcg/kg IV sur 20-30 min (max 20 mcg) – (si répondeurs) *restriction hydrique et suivi Na+ par la suite*	------
Taux inhibiteur HAUT (>5BU)	**Facteur VIIa recombinant activé (rFVIIa)** 90mcg/kg IV en bolus	**Facteur VIIa recombinant activé (rFVIIa)** 90mcg/kg IV en bolus
Thérapie alternative (si facteur non disponible ou comme traitement en cas de haut taux inhibiteur)	**CCP activé (FEIBA)** 85 unités/kg **sinon PCC (Beriplex)** 85 unités/kg (contre-indique si Emicizumab)	------

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:

06/2022

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:

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 units/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

Help Stop the Bleeding

Canadian Hemophilia Society

AHCDC Association of Hemophilia Clinic Directors of Canada

www.hemophilia.ca/emergency