



**NATIONAL HEMOPHILIA FOUNDATION**  
*for all bleeding disorders*

**MASAC Document #257**  
**Replaces Doc #252**

**GUIDELINES FOR EMERGENCY DEPARTMENT MANAGEMENT OF  
INDIVIDUALS WITH HEMOPHILIA AND OTHER BLEEDING DISORDERS**

*The document was approved by the Medical and Scientific Advisory Council (MASAC) of the National Hemophilia Foundation (NHF) on December 2, 2019, and adopted by the NHF Board of Directors on December 5, 2019.*

Patients with bleeding disorders who present to an emergency department for care should receive appropriate, expeditious management. To this end, MASAC has developed the following guidelines.

**Triage**

- 1) Individuals with bleeding disorders should be triaged urgently as delays in administering appropriate therapy, such as infusion of factor concentrate, can significantly affect morbidity and mortality.
- 2) Consultation with the patient's primary provider of bleeding disorder care, in most cases a hematologist, is strongly advised. If this provider is unavailable, consultation with a bleeding disorders provider from the closest hemophilia treatment center is recommended. Administration of clotting factor replacement to the patient should not be delayed waiting for a consultation.

**Assessment**

- 1) Treatment for a suspected bleeding episode is based on clinical history. Physical exam findings may be normal in the early phases of most bleeding episodes associated with an underlying bleeding disorder. Spontaneous bleeding is common in those with severe disease (baseline factor levels <1%). When in doubt, administer clotting factor replacement therapy immediately.
- 2) Treatment decisions should be based on the *suspicion* of a bleeding-related problem, not the documentation of one.
- 3) If the patient or the parent of a patient suspects that occult bleeding is occurring, administer clotting factor replacement. Patients often are instructed to carry with them appropriate factor replacement dosing guidelines as advised by their treating hematologist.

**Diagnostic Studies**

- 1) Clotting factor replacement therapy should be given *before* any diagnostic studies (X-rays, CT scans etc.) are performed to evaluate a suspected bleeding problem, especially in the case of head trauma or suspected intracranial hemorrhage. For routine joint bleeding, *no* radiographic studies are indicated.
- 2) For patients with hemophilia who have illnesses or disorders that necessitate an invasive procedure (lumbar puncture, arterial blood gas, arthrocentesis, etc.) or surgery, factor replacement therapy or bypass therapy to 100% must be administered in the emergency department *prior to* the planned procedure or surgery. In this situation, consultation with a

hematologist is strongly recommended.

- 3) For an individual with known hemophilia, routine laboratory studies (PT, aPTT, factor levels), are not indicated in the treatment of a routine bleeding episode unless requested by the patient's hematologist. Treatment should not be delayed waiting for test results which may take several hours. In some cases, screening assays such as the aPTT and factor activity assays will not be accurate depending on the product the patient is taking and/or the reagents used for the assays at the local laboratory. The clinical severity of a patient's hemophilia is gauged by his or her baseline clotting factor level, a value that remains fairly constant throughout that person's life.

**Indications for Factor Replacement Therapy include:**

- 1) Suspected bleeding into a joint or muscle.
- 2) Any significant injury to the head, neck, mouth or eyes or evidence of bleeding in these areas.
- 3) Any new or unusual headache, particularly one following trauma.
- 4) Severe pain or swelling at any site.
- 5) All open wounds requiring surgical closure, wound adhesive, or steri-strips.
- 6) History of an accident or trauma that might result in internal bleeding.
- 7) Any invasive procedure or surgery.
- 8) Heavy or persistent bleeding from any site.
- 9) Gastrointestinal bleeding leading to moderate to severe anemia.
- 10) Acute fractures, dislocations and sprains.
- 11) Heavy menstrual bleeding leading to moderate to severe anemia or volume instability.

**Treatment**

***Hemophilia A without Inhibitor***

The treatment of choice for individuals with hemophilia A (factor VIII deficiency) is recombinant factor VIII or the patient's product of choice. Plasma-derived concentrate is a suitable alternative in an emergency situation when recombinant factor VIII is not available. Cryoprecipitate and fresh frozen plasma are no longer recommended for treatment of individuals with hemophilia A.

When bleeding is severe, the appropriate dose of factor VIII is **50 units/kg**. This should result in a factor VIII level of 80-100%.

In individuals with hemophilia A receiving prophylaxis with emicizumab who present with acute bleeds, factor VIII should be given, as above. While emicizumab is effective in preventing (prophylaxis) bleeds, it is ineffective in treating acute bleeding events. No adjustment in factor VIII concentrate dosing is recommended for those patients on emicizumab.

***Mild Hemophilia A with Non-Life or Limb Threatening Bleeding***

Individuals with mild hemophilia A (baseline factor VIII greater than 5% and less than 50%) who are experiencing non-life or limb threatening bleeding may respond to desmopressin (DDAVP, see dosing recommendations below). This therapy should be used only if there is documentation in the medical record demonstrating a hemostatic response to this medication. Otherwise, treatment is the same as for other individuals with hemophilia A.

***Hemophilia B without Inhibitor***

The treatment of choice for individuals with hemophilia B (factor IX deficiency) is recombinant factor IX or the patient's product of choice. Plasma-derived concentrate is a suitable alternative in an

emergency situation when recombinant factor IX is not available. Fresh frozen plasma is no longer recommended for treatment of individuals with hemophilia B. Cryoprecipitate does not contain factor IX.

When bleeding is severe, the appropriate dose of factor IX is **100-140 units/kg**. This should result in a factor IX level of 80-100%.

### ***Hemophilia A or B with inhibitor***

**For individuals with inhibitors (antibodies to factor VIII or IX), treatment decisions may be more complicated. The care of inhibitor patients should be urgently discussed with the patient's hematologist. If an individual with an inhibitor presents in a life- or limb threatening scenario, the safest immediate action is to prescribe recombinant factor VIIa (rFVIIa) at a dose of 90 mcg/kg or activated prothrombin complex concentrates (FEIBA) at 75-100 units/kg.\* The patient or family can also provide information on response to these therapeutic bypassing agents.**

**\* Note:** rFVIIa is recommended over FEIBA for acute bleeding events or procedures in hemophilia A inhibitor patients on emicizumab prophylaxis as aPCCs may cause thrombosis or thrombotic microangiopathy in those receiving emicizumab and should be avoided.

**\* Note:** In factor IX patients with a history of inhibitors and anaphylaxis, factor IX-containing products, including FEIBA should be avoided.

### **Additional treatment considerations:**

- 1) If a patient with hemophilia or other bleeding disorder or the parent of a patient with a bleeding disorder brings clotting factor concentrate with them to the emergency department, allow them to utilize it. This recommendation acknowledges many emergency rooms do not have the majority of (or any) clotting factor concentrates on formulary. Patients or family members should be permitted to reconstitute the product and administer it whenever possible. Individuals with bleeding disorders are encouraged to have an emergency dose of factor concentrate or DDAVP (in the form of intranasal Stimate®) in their home and to take it with them when they travel. In those situations when a patient does not bring their own clotting factor concentrate, emergency departments must be prepared to provide clotting factor replacement. Emergency departments must have ready access to factor replacement products so that they are available within one hour of the patient's arrival. In the situation where hospital formulary factor concentrate is used, in order to expedite care, emergency department providers should order **unreconstituted** factor concentrate from their pharmacy or blood bank and reconstitute the product in the emergency department.
- 2) Factor replacement must be administered intravenously by IV push over 1-2 minutes or per label instructions.
- 3) The factor dose should be ordered as "up to the closest vial contents." The full content of each reconstituted vial should be infused, since a moderate excess of factor concentrate will not create a hypercoagulable state but will prolong the therapeutic level of the product administered; thus, it is prudent to "round up."
- 4) When treating an individual with mild hemophilia A who is responsive to DDAVP, the dose and prior responsiveness are usually known. The dose of DDAVP is 0.3 micrograms/kg subcutaneously or intravenously in 30 ml normal saline over 15-30 minutes. It may also be administered as a concentrated nasal spray "Stimate®" at a dose of 1 spray in one nostril for individuals <50 kg and 1 spray in each nostril for individuals >50 kg.
- 5) The most experienced IV therapist or phlebotomist should perform venipunctures. Traumatic venipunctures and repeated needle sticks cause painful hematomas that may limit further IV access.

- 6) In any suspected bleeding emergency in which the clotting factor level of a patient with hemophilia is unknown, the factor level should be assumed to be 0%.
- 7) Intramuscular injections, including immunizations, should be avoided whenever possible. If they must be given, factor replacement therapy must precede the injection. (It can be timed following a prophylactic dose of clotting factor concentrate. This is not required when vaccination is given subcutaneously).
- 8) In situations in which patients are hemodynamically stable and are not requiring volume replacement, the smallest gauge needle should be utilized for obtaining IV access (25g butterfly needles in young infants, 23g butterfly needles in older children and adults).
- 9) Tourniquets should not be applied tightly to extremities because they may cause bleeding.
- 10) Aspirin and aspirin-containing products should be avoided in individuals with hemophilia unless there is a cardiac indication, and then only under close observation for bleeding, e.g. monitoring hemoglobin levels. Acetaminophen and/or oral opioids may be used for analgesia. Non-steroidal anti-inflammatory (NSAID) drugs may be carefully administered to select patients, such as individuals with chronic arthritic pain who are not actively bleeding or being treated for a recent bleeding problem.
- 11) If a patient with hemophilia is bleeding and requires transportation to another facility for definitive care, all efforts should be made to replace the deficient clotting factor **before** transport.

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