NATIONAL HEMOPHILIA FOUNDATION
for all bleeding and clotting disorders

MASAC Document #175

GUIDELINES FOR EMERGENCY DEPARTMENT MANAGEMENT OF INDIVIDUALS WITH HEMOPHILIA

The following guidelines were approved by the Medical and Scientific Advisory Council (MASAC) of the National Hemophilia Foundation on October 14, 2006, and adopted by the NHF Board of Directors on October 15, 2006.

Individuals 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) Triage should be urgent because delays in administering factor concentrate treatment significantly affect morbidity and mortality in individuals with hemophilia.

Assessment

1) Treatment for a suspected bleeding episode is based on clinical history. Physical exam findings tend to be normal in the early phases of most hemophilic bleeds. Spontaneous bleeding is common in individuals with severe disease (factor levels <1%). When in doubt, administer clotting factor replacement therapy.
2) Treatment decisions should be based on the ***suspicion*** of a bleeding-related problem, not the documentation of one.
3) Believe the patient or the parent of a patient. If in their experience they suspect occult bleeding is occurring, administer clotting factor replacement. Patients often are instructed in and/or carry with them appropriate factor replacement dosing guidelines as advised by their treating hematologist.
4) Consultation with the patient's hematologist or a regional hemophilia treatment center professional is strongly advised; however, this should not delay giving clotting factor replacement to the patient.

Diagnostic Studies

1) Clotting factor replacement therapy should be given before any diagnostics studies (X-rays, CAT scans etc.) are performed in the evaluation of 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 must be administered in the emergency department beforehand.
3) For an individual with known hemophilia, routine laboratory studies (PT, PTT, factor levels), are not indicated in the treatment of a routine bleeding episode unless requested by the patient's

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hematologist. 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

1) Suspected bleeding into a joint or muscle.
2) Any significant injury to the head, neck, mouth or eyes or evidence of bleeding in those 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.
10) Acute fractures, dislocations and sprains.

Treatment

Hemophilia A without Inhibitor

The treatment of choice for individuals with hemophilia A (factor VIII deficiency) is recombinant factor VIII or else the patient's product of choice. Plasma-derived concentrate is a suitable alternative in an emergency situation when recombinant Factor VIII is not available. Cryoprecipate 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%.

Mild Hemophilia A with Non-Life or Limb Threatening Bleeding

Individuals with mild hemophilia A (factor VIII greater than 5%) who are experiencing non-life or limb threatening bleeding may respond to desmopressin if they have been shown to respond to this treatment previously. 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 else 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. Note that cryoprecipitate does not contain Factor IX.

When bleeding is severe, the appropriate dose of factor IX is **100-120 units/kg**. This should result in a factor IX level of 80-100%.

1) If a patient with hemophilia or the parent of a patient with hemophilia brings clotting factor with them to the emergency department, allow them to utilize it. They 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 their home and to take it with them when they travel. In those situations where 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 1 hour of the patient's arrival.

In an effort to expedite care, emergency physicians should order unreconstituted factor 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.
3) Dose factor up to the "closest vial" and infuse the full content of each reconstituted vial. A moderate excess of factor concentrate will not create a hypercoaguable state but will prolong the therapeutic level of the product administered; thus it is prudent to “round up.”
4) **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 therapeutic bypassing agents.**
* Note: In factor IX patients with a history of inhibitors and anaphylaxis do not give factor IX-containing products unless the bleeding is life-threatening.
5) When treating an individual with mild hemophilia A who is responsive to desmopressin, the dose and prior responsiveness are usually known. The dose of desmopressin is 0.3mcg/kg subcutaneously or else intravenously in 30 ml normal saline over 30 minutes. It may also be administered as an ultra-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.
6) The most experienced IV therapist or phlebotomist should perform any venipuncture. Traumatic venipunctures and repeated needle sticks cause painful hematomas that may limit further IV access.
7) In any suspected bleeding emergency in which the clotting factor level of an individual with hemophilia is unknown, the factor level should be assumed to be 0%.
8) Intramuscular injections should be avoided if at all possible. If they must be given, factor replacement therapy should precede the injection. Parenteral agents should be given intravenously or subcutaneously. Tetanus immunizations may be administered subcutaneously.
9) 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).
10) Tourniquets should not be applied tightly to extremities because they may cause bleeding.
11) Aspirin and aspirin-containing products are contraindicated in individuals with hemophilia. Acetaminophen and/or codeine may be used for analgesia. Non-steroidal anti-inflammatory 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.
12) If an individual 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.