HEMOPHILIA ASSOCIATION OF NEW JERSEY

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Emergency Guidelines for Hemophilia: General Guidelines

This document was updated and approved July 22, 2016 and originally developed in collaboration with Medical Advisory Board of the Hemophilia Association of New Jersey representatives in November 2005 called Emergency Guidelines for Hemophilia: General Guidelines and from various professional organizations including Medical and Scientific Advisory Council (MASAC) of the National Hemophilia Foundation in October 15, 2006 (document #175) for emergency personnel in New Jersey.

Red Flags in Hemophilia

- All patients are categorized as emergent
- Pain is often the first symptom of bleeding
- All trauma is significant
- **Believe the patient or parent that they are having a bleed.**
- The patient may appear well, yet may be having a significant bleed

1. TREAT HEMOPHILIA PATIENTS IMMEDIATELY.

2. Infuse with Factor BEFORE diagnostic studies, such as radiographs, CAT scans, suturing, or other procedures. For routine joint bleeding, no radiographic studies are indicated.

3. Contact the patient's Hemophilia Treatment Center or hematologist. However, do not delay treatment while waiting for a response. Most patients carry their factor and know their diagnosis or carry an ID card.

4. 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 beforehand.

5. A patient often has pain prior to obvious swelling of a joint. Believe and treat the patient. It is always better to err on the side of treatment rather than withholding factor replacement, since early treatment of joint bleeds can help prevent joint damage.

6. Factor Concentrate must be administered intravenously by IV push over 1-2 minutes.

7. Dose factor up to the "closest vial" and infuse the full content of each reconstituted vial.

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. 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.

10. Intramuscular injections should be avoided if at all possible. If they must be given, factor replacement therapy should precede the injection. Tetanus immunizations may be administered subcutaneously.

11. Tourniquets should not be applied tightly to extremities because they may cause bleeding.

12. Aspirin, NSAIDs and aspirin-containing products are contraindicated in individuals with hemophilia. Acetaminophen and/or codeine may be used for analgesia.

BLEEDS REQUIRING TREATMENT

1. CRITICAL BLEEDS: significant injury to head, neck, face and throat, abdomen, iliopsoas muscle, gastrointestinal bleeding, deep laceration, compartment syndrome, new or unusual headache, severe pain or swelling at any site, acute fracture or dislocations
   Hemophilia A: **Factor VIII** 50 units /kg ***(100 %)***
   Hemophilia B: **Factor IX** 100 -120 units/kg (***100 -120%***)
   von Willebrand Disease: Factor VIII concentrate with von Willebrand factor 50 – 100 units ***VWF: RCo/kg***

   Admit the patient

2. SIGNIFICANT BLEEDS: are bleeding into joint and muscles, soft tissue, sprains, lacerations, hematuria, refractory nosebleeds, oral bleeds
   Hemophilia A: ***Factor VIII*** 15 - 50 units /kg ***(30 -100%)***
   Hemophilia B: ***Factor IX*** 30-100 units /kg ***(30-100%)***

Von Willebrand Disease:

a. Type I DDAVP 0.3 mcg/kg IV drip in 25-100 cc of normal saline (based on total dose of DDAVP) over 30 minutes **maximum dose is 20 micrograms** If not responsive to DDAVP and or for Type 2 A, 2B, and 3
    1. Factor VIII concentrate with von Willebrand factor 50 – 100 units VWF: RCo/kg

Out-patient follow up with hemophilia center should be arranged.

3. Mucosal bleeds, especially the mouth, that spontaneously stop. Usually re-bleed and almost always require treatment with antifibrinolytics agent such as Amicar (amino-caproic acid), 50 mg /kg/dose every 6 hours by mouth to maximum of 12 gm/day, are used in mucosal bleeds to enhance clotting.

RECOMMENDED PRODUCT

The treatment of choice for individuals with Hemophilia A (factor VIII deficiency) or B (factor IX 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. Cryoprecipitate and fresh frozen plasma are no longer recommended for treatment of individuals with hemophilia A or B.

Hemophilia A (Factor VIII Deficiency) Factor VIII Concentrates

Recombinant

Recombinate®
Advate®
Helixate FS ®
Kogenate FS®
Xyntha ®
Novoeight®
Eloctate (Long Acting VIII)

Monoclonal VIII (Plasma Derived)

Hemofil M®
Monoclate P®

Plasma Derived viral inactivated

Alphanate®
Koate DVI®

Hemophilia B (Factor IX Deficiency) Factor IX Concentrates

Recombinant

Alprolix® (Long Acting IX)
BeneFIX®
Rixubus®

Monoclonal IX

Mononine®

Plasma derived viral inactivated

Alphanine

FACTOR VIII INHIBITOR

Recombinate Factor VIIa NovoSeven®

For individuals with inhibitors (antibodies to factor VIII), 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) NovoSeven® at a dose of 90 mcg/kg every 2 hours until bleeding resolves 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.

FACTOR IX INHIBITOR

Do not give factor IX-containing products to patients with a history of factor IX inhibitors and anaphylaxis.

Give Recombinate Factor VIIa. NovoSeven®

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VON WILLEBRAND DISEASE

IV DDAVP

STIMATE INTRANASAL SPRAY

Factor VIII Concentrate containing VWF

Humate P®

Alphanate®

Wilate ®

Koate DVI®

Factor V Deficiencies

Fresh Frozen Plasma

Aminocaproic acid (Amicar)

Factor VII Deficiencies

Recombinant factor VIIa (rFVIIa) NovoSeven® 15-30 mcg/kg every 4 to 6 hours slow IV push

Factor XI Deficiencies

Fresh Frozen Plasma

Aminocaproic acid (Amicar)