Accredo® Specialty Pharmacy

Emergency treatment for patients with bleeding disorders

Immediate steps

This patient needs **immediate attention**—he/she has a bleeding disorder.

+ **Avoid the waiting room and infuse with factor promptly.**
+ **Listen to the patient/caregiver.** He/she understands this condition and the medication.
+ **Contact patient's hematologist immediately.**
  - Hematologist name _____
  - Hematologist phone number _____
+ **Factor infusions** should be administered **BEFORE**: tests, procedures or evaluations—especially with head trauma or suspected intracranial hemorrhage.

Administer factor immediately in these situations

+ Acute fractures, dislocations, sprains or suspected bleeding into a joint or muscle.
+ Any significant injury to the head, neck, mouth or eyes, or evidence of bleeding in those areas.
+ Any new or unusual headache, particularly following trauma.
+ Severe pain, swelling or heavy or persistent bleeding from any site.
+ Open wounds requiring surgical closure, wound adhesive or steri-strips.
+ Gastrointestinal bleeding or history of trauma that may result in internal bleeding.
+ Any invasive procedure or surgery.
+ Heavy or persistent bleeding from any site.

Important facts and tips

+ Limit venipunctures, use of tourniquets and intramuscular (IM) injections.
+ IM injections should be avoided, whenever possible. If required, then factor replacement therapy should precede the IM injection.
+ No radiographic studies needed for routine joint bleeding.
+ Factor dosage is based on the patient's weight (units/kg).
+ If the circulating factor level of the patient is unknown, then assume it to be 0%.
+ Most reconstituted factor can be infused by slow IV push over a short period of time. Each product may vary slightly based upon the volume of drug prescribed. Check product package insert (PI) for guidance.
+ Side effects including tingling at the site (or in the hands or lips), lightheadedness, shortness of breath and burning at the infusion site may occur.
+ Gross hematuria in bleeding disorder patient—DO NOT treat with factor until speaking with his/her hematologist.
+ Patients may have brought factor with them. In order to avoid delaying treatment, allow them to reconstitute and administer, if necessary.

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Recommended bleeding disorder treatments

Treatment of hemophilia A (factor VIII deficiency)

1. Recombinant factor VIII concentrates
2. Plasma-derived factor VIII concentrates
3. Cryoprecipitate not recommended
4. Treatment of mild hemophilia A: Desmopressin nasal spray or injection (Stimate® or DDAVP® injection). If not responsive to desmopressin, factor VIII concentrate is recommended.

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%. For dosing confirmation, see medication PI as this may vary by product.

Treatment of hemophilia B (factor IX deficiency)

1. Recombinant factor IX concentrate
2. Plasma-derived factor IX concentrates
3. Avoid protein complex concentrates (Bebulin®, Profilnine®) to limit thromboembolic risk.

When bleeding is severe, the appropriate dose of factor IX is approximately 100 units/kg. This should result in a factor IX level of 80–100%. For dosing confirmation, see medication PI as this may vary by product.

Treatment of hemophilia A and B with inhibitors

Dosing best determined through immediate discussion with patient's hematologist.

1. Factor Eight Inhibitor Bypassing Activity FEIBA®, anti-inhibitor coagulant complex—intravenous infusion rate not to exceed 2 units/kg/min.
2. NovoSeven® RT, coagulation factor VIIa—the preferred agent for Factor IX patients with a history of inhibitors.

Treatment of von Willebrand (vWD)

See medication PI for dosing recommendations as this may vary by product.

Type 1: Treat with the synthetic intravenous desmopressin or Stimate® 1.5 mg/mL nasal spray for bleeding.

Type 2A, 2M and 2N: Consider treating with DDAVP, if patient previously showed a response to a DDAVP trial (per patient history). If patient has already treated with DDAVP or Stimate prior to ER visit, then a product containing von Willebrand factor (vWF) such as Alphanate®, Humate-P®, Koate®*, Vonvendi® and Wilate® should be considered.

Type 2B and type 3 – and type 1, 2A, 2M and 2N nonresponsive to DDAVP: Treat with a product that contains the vWF factor protein such as Alphanate®, Humate-P®, Koate®*, Vonvendi® and Wilate®.

We're here to help 24/7/365

Contact a clinician with access to this patient's chart at Accredo specialty pharmacy—866.712.5007.

Medical and Scientific Advisory Council (MASAC)—National Hemophilia Foundation. https://www.hemophilia.org. Accessed Nov. 21, 2016.

*Koate® is a suggested option per MASAC guidelines but not FDA-approved for the treatment of von Willebrand disease.

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