SECTION 8B: PUMPS & DRUG ADMINISTRATION	POLICY: 8A.8
POLICY: COAGULANT BLOOD PRODUCTS FOR BLEEDING DISORDERS	PAGE: 1 OF: 10

PURPOSE:

To provide instruction on the safe and proper administration of Factor medications and similar blood products for the treatment of bleeding disorders to patients in the home setting.

POLICY:

Factor medications and similar blood products (listed) will be administered by a trained RN in accordance with a physician's order, any available / provided Pharmacy or manufacturer instructions, and Agency policy.

- Advate
- Adynovate
- Alphanate
- Aprolix
- Benefix
- Eloctate
- Helixate FS
- Hemlibra
- Hemofil
- Humate-P
- Idelvion
- Kogenate FS
- Novoseven
- Novoeight
- Recombinate
- Von Vendi
- Xyntha

GENERAL INFORMATION:

- Bleeding disorders are a group of disorders that share the inability to form a proper blood clot. They are characterized by extended bleeding after injury, surgery, trauma, or menstruation. Sometimes the bleeding is spontaneous, without a known or identifiable cause. Improper clotting can be caused by defects in blood components such as platelets and/or clotting proteins, also called clotting factors.

- The body produces 13 clotting factors. If any of them are defective or deficient, blood clotting is affected; a mild, moderate, or severe bleeding disorder can result.

- Treatment for bleeding disorders varies, depending on the condition and its severity. For some bleeding disorders, there are clotting factor concentrates that can be infused prophylactically or on-demand at home, to prevent or treat bleeds. For other bleeding disorders, there are topical products, nasal sprays, and fresh frozen plasma, which is administered in a hospital setting.

- Bleeding disorders can be inherited or acquired from such conditions as anemia, cirrhosis of the liver, HIV, leukemia, and vitamin K deficiency. They also can result from certain medications that thin the blood, including Aspirin, Heparin and Warfarin.

- Hemophilia is an inherited (genetic) disorder in which the blood doesn't clot normally because it lacks sufficient blood-clotting proteins (clotting factors). If a person has hemophilia, they may bleed for a longer time after an injury than would if their blood clotted normally.

- Small cuts usually aren't much of a problem. The greater health concern is deep bleeding inside the body, especially in the knees, ankles, and elbows. That internal bleeding can damage the organs and tissues and may be life-threatening.

SYMPTOMS

- Symptoms of a bleeding disorder include:
    - Bleeding into joints, muscles, and soft tissues
    - Excessive bruising
    - Prolonged, heavy menstrual periods (menorrhagia)
    - Unexplained nosebleeds
    - Extended bleeding after minor cuts, blood draws or vaccinations, minor surgery, or dental procedures

- Signs and symptoms of hemophilia vary, depending on the level of clotting factors. If the clotting-factor level is mildly reduced, patient may bleed only after surgery or trauma. If deficiency is severe, they may experience spontaneous bleeding.

- Signs and symptoms of spontaneous bleeding include:
    - Unexplained and excessive bleeding from cuts or injuries, or after surgery or dental work.
    - Many large or deep bruises.
    - Unusual bleeding after vaccinations.
    - Pain, swelling or tightness in your joints.
    - Blood in your urine or stool.
    - Nosebleeds without a known cause.
    - In infants, unexplained irritability.

- A simple bump on the head can cause bleeding into the brain for some people who have severe hemophilia. This rarely happens, but it's one of the most serious complications that can occur. Signs and symptoms include:
    - Painful, prolonged headache.
    - Repeated vomiting.
    - Sleepiness or lethargy.
    - Double vision.
    - Sudden weakness or clumsiness.
    - Convulsions or seizures.

TYPES OF BLEEDING DISORDERS:

- Hemophilia A (Factor VIII deficiency)

  Hemophilia A, also called factor VIII (FVIII) deficiency or classic hemophilia, is a genetic disorder caused by missing or defective factor VIII, a clotting protein. Although it is passed down from parents to children, about 1/3 of cases are caused by a spontaneous mutation, a change in a gene.

  According to the US Centers for Disease Control and Prevention, hemophilia occurs in approximately 1 in 5,000 live births. There are about 20,000 people with hemophilia in the US. All races and ethnic groups are affected. Hemophilia A is four times as common as hemophilia B while more than half of patients with hemophilia A have the severe form of hemophilia.

- Hemophilia B (Factor IX deficiency)

  Hemophilia B, also called factor IX (FIX) deficiency or Christmas disease, is a genetic disorder caused by missing or defective factor IX, a clotting protein. Although it is passed down from parents to children, about 1/3 of cases are caused by a spontaneous mutation, a change in a gene.

According to the US Centers for Disease Control and Prevention, hemophilia occurs in approximately 1 in 5,000 live births. There are about 20,000 people with hemophilia in the US. All races and ethnic groups are affected. Hemophilia B is four times less common than hemophilia A.

- Von Willebrand Disease

  Von Willebrand disease (VWD) is a genetic disorder caused by missing or defective von Willebrand factor (VWF), a clotting protein. VWF binds factor VIII, a key clotting protein, and platelets in blood vessel walls, which help form a platelet plug during the clotting process. The condition is named after Finnish physician Erik von Willebrand, who first described it in the 1920s.

  VWD is the most common bleeding disorder, affecting up to 1% of the US population. It is carried on chromosome 12 and occurs equally in men and women.

- Rare Factor Deficiencies (I, II, V, VII, X, XI, XII and XIII.)

  In the US, a rare disease or disorder is defined as one that affects fewer than 200,000 people, making hemophilia A and B, and still less prevalent factor deficiencies such as I, II, V, VII, X, XI, XII and XIII, rare disorders. These very rare factor deficiencies, from factor XIII deficiency, the rarest, occurring in an estimated 1 out of 5 million people, to factor XI deficiency, occurring in about 1 out of 100,000, were all discovered and identified in the 20th century. Most of these conditions were only identified within the last 60-70 years.

INHIBITORS & COMPLICATIONS:

- **Adverse reaction to clotting factor treatment.** Some patients develop an immune response to the medications used to treat hemophilia. The immune system is the primary defense system of the body against disease and foreign agents. For these patients, the immune system produces antibodies that “inhibit” clot formation by destroying the clotting factor before it has a chance to stop the bleeding. The reason for a patient developing inhibitors is still not entirely clear and the presence of an inhibitor makes treating bleeds more difficult. Standard treatment does not work, and other methods to control bleeding must be used.

- **Unusual Bleeds**. Bleeds beneath the skin can cause large bruises that take time to heal. Bleeding in certain areas—the eye, head, brain, throat, or gastrointestinal tract—can be life threatening and require immediate attention.

- **Damage to joints.** Bleeding into joints and muscles can cause swelling, pain, and immobility. Bleeds in muscles can cause permanent nerve damage when the trapped blood or swollen muscles put pressure on a nerve. Some sites can become target joints, where repeat bleeds damage the lining of the joint, called the synovium. Synovitis is a condition where chronic inflammation in the joint from recurrent bleeds has thickened the synovium, reducing the joint space and limiting movement. The bones can then become arthritic and develop cysts. Eventually, there may be so much damage that a joint replacement or fusion may be needed. Deep internal bleeding. Bleeding that occurs in deep muscle can cause the limbs to swell. The swelling may press on nerves and lead to numbness or pain.

- **Infection.** People with hemophilia are more likely to have blood transfusions, increasing their risk of receiving contaminated blood products. Blood products became safer after the mid-1980s due to screening of donated blood for hepatitis and HIV.

SPECIAL CONSIDERATIONS: HOME THERAPY

- Where appropriate and possible, persons with hemophilia should be managed in a home therapy setting.

- Home therapy allows immediate access to clotting factor and hence optimal early treatment, resulting in decreased pain, dysfunction, and long-term disability and significantly decreased hospital admissions for complications.

- Further improvements in quality of life include greater freedom to travel and participate in physical activities, less absenteeism, and greater employment stability.

- Home therapy is ideally achieved with clotting factor concentrates or other lyophilized products that are safe, can be stored in a domestic fridge, and are reconstituted easily.

- Home treatment must be supervised closely by the comprehensive care team and should only be initiated after adequate education and training.

- **Teaching** should focus on general knowledge of hemophilia; recognition of bleeds and common complications, first aid measures, dosage calculation, preparation, storage, administration of clotting factor concentrates, aseptic techniques, performing venipuncture (or access of central venous catheter), record keeping, proper storage and disposal of needles/sharps, and handling of blood spills. RN should assess the patient's/family's understanding of the specific coagulation disorder, its management and potential complications as well as determine educational, support, and treatment follow-up needs. Consideration should be given to the patient's developmental stage, cultural and educational background, and individual patient and family characteristics that might impact on their understanding.
    - Agency shall monitor patient/family for no less than one (1) month and two (2) demonstrated infusions following administration independence.
- Patients or parents should keep bleed records (paper or electronic) that include date and site of bleeding, severity and length of bleed, dosage and lot number of products used, and adverse effects.
- Patients or parents should be instructed to keep their doctor’s emergency contact information readily available and always keep a diagnosis letter from their doctor with them.
- Home care can be started with young children with adequate venous access and motivated family members who have undergone adequate training.
- Older children and teenagers can learn self-infusion with family support.
- An implanted venous access device (Port-A-Cath) can make injections much easier and may be required for administering prophylaxis in younger children. The venous access device must be kept scrupulously clean and be flushed monthly, at a minimum, and after each administration to prevent clot formation.

SPECIAL CONSIDERATIONS: ADMINISTRATION

- In addition to training on bleeding disorders and standard procedures and protocols, RNs administering factor medications shall be trained on each medication and delivery system they are tasked with administering.
    - Individual administration guides and videos for these drugs are the primary and reliant source of documentation for step-by-step administration instructions.

- Most medications are administered intravenously via a slow IVP.
    - IV access shall be maintained and assessed per agency policy and protocols.
    - Site(s) should be rotated with each administration.
    - Monitor for bleeding after administration.
    - If bleeding occurs, hold pressure until bleeding stops.

- **Hemlibra**, unlike the other drugs, is administered via sub-cutaneous injection.

- Severe hemophilia patients are treated prophylactically for bleeds, up to three (3) times weekly.

- For slow IVP administration of a prophylactic dose, a butterfly needle is most common and appropriate.
    - Prime the butterfly needle with saline. (If pharmacy does not send saline, please reach out to Agency as we will have to contact Pharmacy to get approval to access patient prior to mixing medication.)
    - Once butterfly is in the vein there will be blood return.
    - Secure butterfly needle in place with tape.
    - Mix factor medications (**Do Not** tamper with medication until access is established).
    - Twist off saline syringe and place factor syringe on tubing.
    - Slowly infuse.
    - When complete, twist off factor syringe, and use saline syringe to flush all remaining medication within the tubing.
    - Remove butterfly needle.
    - Check for bleeding from site and hold pressure if bleeding occurs.
    - **Document** the procedure in its entirety including any complications. Be sure to record the expiration date(s) and lot number(s) for all vials used.

- For spontaneous bleeds and PRN dosing:
    - RN must ensure catheter access, catheter assessment, and bleed assessment is completed no less than daily. Families educated on medication administration and catheter maintenance may self-administer during spontaneous bleeds.
    - Bleed needs to be treated as ordered (every 12-24 hours per order) until the bleed is completely resolved.
    - IVs can be left in place or ports can stay accessed per protocol for the duration of the bleed.
    - Agency shall provide patient & family with education on how to identify bleeds and contact Agency for nursing.
    - Agency shall provide patient & family with 24-hr on-call phone/text line, along with contact information for primary RN and Director of Nursing.
    - Agency shall make all efforts to administer PRN dosing within two (2) hours of reported bleeds.
    - If/when the Agency is unable to accommodate PRN dosing in a timely manner (based on severity of bleed), patient shall be encouraged to seek care at the nearest emergency room.