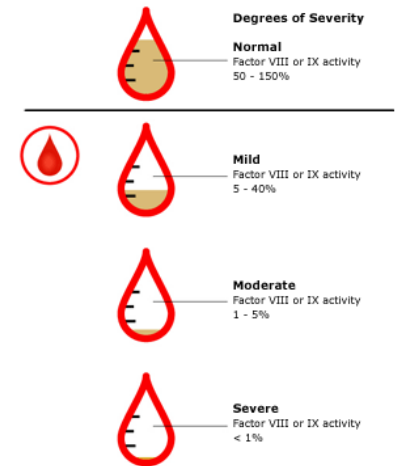


What is it?

Hemophilia is a hereditary disorder affecting the blood's ability to coagulate, or clot. Hemophilia is recessive X linked trait; females are carriers and males most often affected. However, in some cases there is no family history and it is believed the disorder is caused by a new gene mutation.

Children with hemophilia have a deficient or absent protein (factor) found in plasma that is needed to prevent excessive bleeding. The most common missing clotting factors are factor VIII (Hemophilia A) and factor IX (Hemophilia B). Hemophilia severity ranges from mild to severe in proportion to the actual clotting factor deficiency.




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What are the symptoms?

Symptoms of hemophilia range depending on the injury. Injuries can result from trauma or spontaneous bleeding. Affected children bleed excessive, uncontrolled amounts *longer*, not faster. Not all bleeds are visibly seen as they most commonly occur in joints and muscles. Symptoms can include bruising, swelling, redness and pain. A child may feel the symptoms of their affected "target joint" long before signs of a bleed are visible.

Types of Bleeds

Joint

- Early sign "tingly and warm"
- Pain, swelling, hard to move
- Ankles, knees, elbows

Muscle

- Tight skin, stiff, swollen
- Very painful
- May bruise
- Calf, thigh, forearm

Soft tissue and Minor cuts

- Bruising, hematoma visible
- Monitor time to stop bleeding
- Bleeding usually stops with RICE method

Life threatening bleeds require *immediate* attention because of their location and their potential to bleed into an enclosed space and press on vital tissue.

Head– nausea, vomiting, headache, confusion, change in LOC

Spine– back pain, tingling, numbness

Throat– difficulty swallowing, breathing

Abdomen– pain, tenderness, swelling, bloody stools

Ocular– pain, change in vision

What is the treatment?

- Early and appropriate treatment when bleeds occur is critical to prevent complications. More severe bleeding and bleeding into muscles, joints, or internal organs must be treated by giving replacement factor immediately. Replacement of the deficient clotting factor concentrate is delivered directly into the vein, via a peripheral IV stick or central line or port. This treatment is called *episodic or on-demand concentrates*. Some children will also be on a prophylactic treatment where they take medication on a regular schedule to reduce the risk of spontaneous bleeds.
- Other medications include therapies that are *non-factor* based. Antifibrinolytics prevent the breakdown of blood clots by neutralizing chemicals in the blood and mucous membranes (nose, mouth, urinary tract). Brand names include Amicar (used for nose/mouth bleeds) and Lysteda (used for heavy menstrual flow). Another *non-factor* therapy is Desmopressin Acetate, which works by affecting a protein in the blood which in turn can raise the blood clotting factor necessary for clotting. Brand names include DDAVP and Stimate nasal spray.

SHNIC school nurses information:

Specific health issues for individual health care plans

- Diagnosis type and severity of hemophilia
- Contact information for caregiver, emergency department and local hemophilia treatment center (HTC)
- Most affected areas with signs and symptoms, frequency of bleeds
- Child's ability to report symptoms
- Hemophilia emergencies, hospitalizations and ER visits
- Medications and schedule for prophylactic treatment, pain
- PRN pain medications
- Orders for vascular device like central line or port, if applicable with emergency plan
- When to initiate treatment and/or access vascular device
- Factor concentrate medication to be given at school, if ordered
 - ⇒ Brand name, dose, frequency, indications
 - ⇒ Storage and supplies needed
 - ⇒ Preparation and method of administration
- Monitoring for fever if central line accessed, when to notify healthcare provider
- Physical activity restrictions
- Re-evaluation requirements by MD before returning to activities after a bleeding episode
- Medical alert bracelet

Resources & Manuals

Partners in bleeding disorders education: Education for all healthcare providers

<http://partnerspm.org/>

Hemophilia Federation of America

<http://www.hemophiliafed.org/>

Playing it safe- Bleeding disorders sports and exercise

<http://www.hemophilia.org/sites/default/files/document/files/PlayingItSafe.pdf>

Dads in Action: Students with bleeding disorders

http://www.rarebleedingdisorders.com/content/dam/medical/rarebleedingdisorders/resources/documents/for-caregivers-and-parents/Dads%20in%20Action_Students%20with%20Bleeding%20Disorders.pdf