HEMOPHILIA

What is it?

Hemophilia is a hereditary bleeding disorder affecting the blood's ability to coagulate or clot. People with this condition experience prolonged bleeding or oozing following an injury, surgery, or procedure. Bleeding may also start again several days post injury or surgery. Children with hemophilia have a deficient or absent protein (factor) in plasma necessary 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 clotting factor deficiency. Hemophilia is a recessive X-linked trait meaning males are most often affected. However, in some cases there is no family history and researchers believe the disorder is caused by a genetic mutation.

Hemophilia can result in bleeding within joints, head, and brain. Individuals with hemophilia bleed excessive and uncontrolled amounts of blood *longer,* not faster.

What are the characteristics or complications?

Not all bleeds are visible. Bleeds can occur internally into muscles and joints or externally from minor cuts, dental procedures, or trauma. Bleeds that occur without obvious cause are called spontaneous bleeding episodes. Hallmark symptoms include unexplained or excessive bleeding, bruising, swelling, redness, and pain. A child may feel the symptoms of their affected "target joint" long before signs of a bleed are visible.

- Frequent nose bleeds
- Bleeding gums
- Blood in urine or stool
- Vomiting blood
- Joint pain or swelling
- Large and deep bruises
- Irritability

Life threatening bleeds require *immediate* attention because of their location and their potential to bleed into an enclosed space pressing 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

Kennedy Krieger Institute

The **Specialized Health Needs Interagency Collaboration (SHNIC)** program is a collaborative partnership between the Kennedy Krieger Institute and the Maryland State Department of Education.

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 immediately by administering replacement factor. Replacement of the deficient clotting factor concentrate is delivered directly into the vein via a peripheral IV stick, 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). Another non-factor therapy works by affecting a protein in the blood that can raise the blood clotting factor necessary for clotting.

Suggested school accommodations

School personnel should be educated about hemophilia and understand the student emergency plans including an Emergency Evacuation Plan (EEP). A 504 plan may also be considered.

Specific health issues for Individualized Healthcare Plan

- 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
- Student's ability to report symptoms
- History of hemophilia emergencies, hospitalizations, and ER visits
- Medications and schedule for prophylactic treatment and pain medications including factor concentrate medication to be given at school, if ordered
  - Brand name, dose, frequency, indications
  - Storage and supplies needed
  - Preparation and method of administration
- Orders for vascular device like central line or port, if applicable and when to access
- Monitoring for fever if central line accessed and when to notify healthcare provider
- Physical activity restrictions
- Re-evaluation requirements before returning to activities after a bleeding episode
- Medical alert bracelet
- Emergency Care Plan (ECP)

Resources & Manuals

National Hemophilia Foundation
https://www.hemophilia.org/

Hemophilia Federation of America
https://www.hemophiliafed.org/

Partners in Bleeding Disorders Education
https://partnersprn.org/