

HEMOPHILIA

Background

Hemophilia is a hereditary bleeding disorder affecting the blood's ability to coagulate or clot. Bleeds can occur externally from minor cuts or injury or internally into muscles and joints.

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.

The most common type of bleeds occurs within joints and muscles and requires immediate attention. Spontaneous bleeding episodes can occur in both locations; meaning the bleed occurs without any known trauma. Early joint bleeds are characterized by tingling, pain, and limited range of motion before progressing to heat, swelling, and further decreased range of motion. Muscle bleeds are characterized by redness, swelling, pain and heat. Any injury to the head, throat, abdomen, or eyes is considered an emergency because of their location and their potential to bleed into an enclosed space pressing on vital tissue. Signs and symptoms of these areas that require immediate attention could include:

- Nausea, vomiting
- Headache
- Confusion
- Difficulty swallowing
- Abdominal pain, tenderness
- Bloody stool
- Eye pain, change in vision
- Difficulty breathing
- Change in level of consciousness

Early and appropriate intervention 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 using medication to reduce the risk of spontaneous bleeds.

Top Takeaways for School Considerations

A student diagnosed with hemophilia has prolonged bleeding time. Individuals with hemophilia bleed excessive and uncontrolled amounts of blood longer, not faster.

Bleeding can occur as the result of an injury or spontaneously. Bleeding into the muscles, joints, or internal organs can be life threatening.

School personnel should be educated about the student's type and severity of hemophilia and understand the student's emergency plan. School personnel should be aware of student's ability to self-report symptoms of a bleed.

Pain or injury should be reported to the school nurse. If the student reports pain, even without visible injury, treatment may be needed. Contact information for the local hemophilia treatment center (HTC) is important.

The student may have a vascular access device (e.g., port or central line) to administer emergency medication.

Kennedy Krieger Institute's Specialized Health Needs Interagency Collaboration

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



Kennedy Krieger Institute

Considerations for the Individualized Healthcare Plan (IHP)

- Nursing diagnosis of risk for injury, impaired physical mobility and pain (acute/chronic)
- Current diagnosed health condition including date of diagnosis, progress of disease process and other chronic health conditions
- Current medication and treatment orders (consider schedule, equipment needs and side effects)
- Assessment of implanted medical device (consider location, date of surgical placement, and device specific information)
- Activity, positioning, transferring (consider precautions and/or restrictions such as re-evaluation before returning to activities after a bleeding episode)
- Use of specialized equipment, adaptive equipment, and orthotics
- Consider emergency care plan(s) (ECP) and emergency evacuation plan(s) (EEP) as related to medical needs in the school setting, and staff education/training, as appropriate

Discussion Starters for Educational Team

1. Has the school staff been trained in implementing the student-specific emergency plan as applicable?
2. Would the student benefit from evaluations or assessments in any of the following areas: physical therapy, occupational therapy, speech and language therapy, assistive technology, adapted physical education, functional behavior, psychology, hearing and vision?
3. Would the student benefit from additional academic support and/or modified education (e.g., copies of notes, extra time, reduced workload, simplified instructions, alternative formats for presentation of material, 504/IEP)?
4. Does the student require activity precautions to prevent injury?
5. Is the physical school environment safely accessible for the student's mobility needs (e.g., entry and exit, ramps, location of classes, access to elevator, doorways)?

Resources

Hemophilia Federation of America
hemophiliated.org/

Partners in Bleeding Disorders Education
partnersprn.org/

Resources National Hemophilia Foundation
hemophilia.org/



Scan QR code or visit **KennedyKrieger.org/Redirect** for more information.