

Bleeding Disorders: An Overview

What are bleeding disorders?

- *Bleeding disorders* is a general term used to describe medical conditions in which the blood does not clot well.
- The process of blood clotting is very complex, and things can go wrong at many stages. Two parts of the blood are required for effective clotting: tiny cells called *platelets* and proteins called *clotting factors*. When a child's blood does not clot, the child may bruise easily, have nosebleeds, or bleed for a long time after being injured or after undergoing surgery.
- Platelet problems are a type of bleeding disorder.
 - Having a low platelet count can interfere with normal clotting. Low platelet counts can be caused by different diseases or be a side effect of certain medications.
 - The most common disease that causes low platelet counts is immune thrombocytopenia. See Immune Thrombocytopenia Quick Reference Sheet (page 153) for more details.
 - Also possible are rare disorders of platelet function that can result in similar bleeding tendencies.
- Clotting factor disorders are another type of bleeding disorder.
 - The 2 most common of these disorders are von Willebrand disease and hemophilia.
 - Von Willebrand disease can affect males and females equally, but hemophilia usually affects only males.

How common are they?

- Statistics about von Willebrand disease vary, but it probably affects about 1% to 2% of the US population.
- Hemophilia occurs in 1 in 5,000 boys at birth and affects approximately 20,000 people, mostly males, in the United States.

What are some common characteristics of children who have bleeding disorders or of bleeding disorders as children present with them?

- Bleeding disorders may cause lumps under the skin (called *hematomas*); or flat, patchy collections of bleeding (called *purpura*); or pinpoint bleeding in the skin (called *petechiae*).
- Spontaneous bleeding or bleeding after injury can occur in the joints or the gastrointestinal and urinary tracts. Joint bleeding can occur in children with hemophilia and is very painful.

- Nosebleeds and bleeding from the gums or teeth may also be common.
- Hemophilia is characterized as follows:
 - Spontaneous, deep, and excessive surgical bleeding is more common in children with hemophilia. Bleeding into the joints is very painful and causes problems over time; efforts are made to prevent this complication.
 - Children with hemophilia are often treated with infusion of the missing clotting factors. In the past, these clotting factors were derived from blood and placed children with hemophilia at risk for blood-borne diseases and infections such as hepatitis B and C and HIV infection, but changes in the way clotting factors are produced have reduced this problem. Since the middle to late 1980s, treatment of these clotting factors, as well as the use of genetically engineered clotting factors, has essentially eliminated the risk of blood-borne diseases and infections.
- Von Willebrand disease is characterized as follows:
 - Children with it usually present with more superficial bleeding, such as from the nose, from the mouth, and menstrual bleeding.
 - It can vary in severity, but most forms cause less-serious bleeding than hemophilia.
 - Some children with mild von Willebrand disease do not need any special adaptations, just awareness of their conditions in case they are injured or require emergency treatment.
 - Treatment can include any, or any combination of, clotting factor replacement products, desmopressin acetate, and e-aminocaproic acid (Amicar) or tranexamic acid.

Who might be on the treatment team?

- Children with hemophilia and children with von Willebrand disease require specialty care. The doctors who care for them are called *hematologists*, and they treat them along with specialty nurses, physical therapists, and social workers, as well as the pediatrician/primary care provider in the medical home.
- Children with von Willebrand disease bleed less than children with hemophilia and therefore need less urgent and frequent specialty care, but hematologists are usually involved in the care of these children and their bleeding problems prior to surgery or procedures or after trauma.

Bleeding Disorders: An Overview *(continued)*

What adaptations may be needed?

Bleeding disorders vary in types and severity, so it is best to obtain details about a specific child's needs from parents/guardians and the child's specialty doctors. At a minimum, a high index of suspicion for the possibility of bleeding, even with mild trauma, is needed. Also, any signs or symptoms of bleeding should initiate a rapid appropriate response (as indicated in the individual child's treatment plan) to control bleeding and minimize complications.

Medications

- A medication called *desmopressin acetate* can help prevent or eliminate bleeding caused by some types of bleeding disorders. It can be injected under the skin or sprayed into the nose. In medical settings, it can be given intravenously.
- Some children with a more severe form of hemophilia will have a long-term intravenous catheter placed so they can be treated with clotting factor replacement products more easily.
- Amicar and tranexamic acid are useful supportive medications for patients with hemophilia and von Willebrand disease. Once a clot is formed, these medications help stabilize the clot by inhibiting the body's natural process for breaking it down. This allows more time for a wound to heal without rebleeding. These medications are commonly used when patients have dental procedures and for control of excessive menstrual bleeding. Caution is required when deciding to use these medications if bleeding involves a location where a stable clot may cause additional damage/injury, such as the upper urinary tract (ie, kidney), the cranium (subdural hematoma), or a joint.
- All children with clotting factor disorders should avoid nonsteroidal anti-inflammatory drugs such as ibuprofen (eg, Advil, Motrin), naproxen (eg, Aleve), and aspirin.
- Acetaminophen (eg, Tylenol) is usually fine to use, but patients with bleeding disorders should discuss any medications to be taken, even over-the-counter medications, with their respective hematologists.
- All staff who will be administering medication should have medication administration training (see Chapter 6).

Dietary Considerations

There is no special diet for bleeding disorders, but it is important that the child not develop overweight, because that condition can put more stress on the joints.

Physical Environment and Other Considerations

- Preventing trauma, especially to the head, can be a big challenge, particularly in the preschool years. Be extra cautious that straps in high chairs are fastened and that children are watched carefully on elevated surfaces such as changing tables and when climbing. Pad any sharp corners. Some children with severe hemophilia may wear helmets to protect their heads, but this precaution is not common and is usually done to reduce the risk of further bleeding in a child who already experienced head bleeding. Gym activities may need to be adapted for school-aged children.
- First aid consists of cool compresses that can be used for bleeding. Popsicles are sometimes helpful for mouth injuries, but do not allow the child to keep the popsicle in one place for too long, to prevent cold injury.
- If a nosebleed occurs, pinch the end of the nose below the nasal bone for 10 minutes and have the child stay in a neutral position.
- Exercises such as bicycle riding, walking, and swimming are good ways to keep muscles strong and joints flexible. Have the child wear appropriate safety equipment (eg, helmets, elbow pads, kneepads).
- Use standard precautions when dealing with bleeding.

What should be considered an emergency?

- Call emergency medical services (911) for
 - Head trauma followed by headache, vomiting, change in behavior, or another unusual sign
 - Any bleeding that is not easily stopped after 10 minutes, or any vigorous bleeding
- Call parents/guardians for
 - Swelling of a joint or muscle. Parents/guardians should be notified immediately about any signs or symptoms of bleeding into a muscle or joint, such as swelling or inability to move the body part. Older children may recognize the sensation of a joint with bleeding or they may notice swelling of their muscles or joints. You should notify parents/guardians immediately.
 - Minor episodes of bleeding that are stopped with first aid.

Bleeding Disorders: An Overview *(continued)*

What types of training or policies are advised?

- First aid for bleeding
- Standard precautions such as using gloves, washing hands, and sanitizing surfaces when dealing with blood (See Glossary for further details.)
- Background on hemophilia or von Willebrand disease

What are some related Quick Reference Sheets?

Immune Thrombocytopenia (page 153)

What are some resources?

- National Heart, Lung, and Blood Institute: www.nhlbi.nih.gov, 301/592-8573—“What is hemophilia?” (Web page), www.nhlbi.nih.gov/health/health-topics/topics/hemophilia
- National Hemophilia Foundation: www.hemophilia.org, 212/328-3700
 - “Playing It Safe for Kids” (in-person workshop), www.hemophilia.org/Events-Educational-Programs/In-Person-Workshops/Playing-it-Safe-for-Kids
 - Steps for Living (Web site), <https://stepsforliving.hemophilia.org>

