

Quick Facts

What are the most common types of bleeding disorders?

- Hemophilia A (factor VIII [8] deficiency)
- Hemophilia B (factor IX [9] deficiency)
- von Willebrand disease (VWD)

What is hemophilia?

- Hemophilia is an inherited bleeding disorder in which there is a deficiency or lack of factor (most commonly VIII (8) (hemophilia A) or factor IX (9) (hemophilia B) in a person's blood.
- Hemophilia A or B can be mild, moderate, or severe.

What is VWD?

- VWD is a bleeding disorder caused by a defect or deficiency of a blood clotting protein, called von Willebrand factor (vWF).
 - vWF is a protein critical to the initial stages of blood clotting. This glue-like protein, produced by the cells that line the blood vessel walls, interacts with blood cells called *platelets* to form a plug, which prevents the blood from flowing at the site of injury.
 - People with VWD are unable to make this plug because they do not have enough vWF or their factor is abnormal.
- VWD is a genetic disorder that can be inherited from either parent.
- It can affect males and females equally.
 - VWD can be Type 1, Type 2a, Type 2b, Type 3.
 - Type 1 is typically the mildest form, while Type 3 is more severe.

What is the incidence of VWD?

- VWD is estimated to be present in 1% to 2% of the population.

What is the incidence of hemophilia?

- Hemophilia A: 1 in 5,000 male births
- Hemophilia B: 1 in 30,000 male births



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Quick Facts (continued)

What types of bleeding should I expect in a person with a bleeding disorder?

- Joint bleeding (internal bleeding into the joints)
 - Symptoms:
 - Tingling or bubbling sensation
 - Stiffness
 - Warmth
 - Pain
 - Unusual limb positions
- Muscle bleeding (internal bleeding into the muscles)
 - Symptoms:
 - Same as joint bleeding.
- Soft tissue bleeding (internal bleeding marked by excessive bruising or swelling)
 - Life-threatening bleeding (head injuries, eyes, neck/throat, abdominal/stomach, kidney/bladder)
 - Other common bleeding (nosebleeds, mouth/teeth)

How are bleeding disorders treated?

- Bleed disorders are treated with replacement of missing clotting proteins via intravenous infusion.
- This can be done
 - On demand (as injuries/bleeds occur on an as needed basis)
 - Prophylaxis (a preventative treatment program given regularly to prevent bleeds before they happen)
- Many parents and even older children have learned to infuse factor at home either through a venous access device (a port-a-cath or med-a-port) or peripherally (directly into a vein).
- Persons with Von Willebrand Disease may use drugs also given intravenously or via a nasal spray.

How do I help someone who is having a bleed?

- Believe a person when he/she says something hurts.
- Use standard first aid for scrapes, scratches, and minor cuts.
- Use R.I.C.E. therapy: Rest, Ice, Compression, Elevation.
- Begin factor replacement therapy as soon as a bleed is detected!

What happens if a person with a bleeding disorder is left untreated?

- Continued or repeated bleeding into the joints can cause:
 - Flexion contractures
 - Joint arthritis/arthropathy
 - Chronic pain
 - Muscle atrophy
- Continued or repeated bleeding into muscle can cause:
 - Compartment syndrome
 - Neurologic impairment