hemophilia?

Mild hemophilia is an inherited disorder that results in prolonged or delayed bleeding with tooth extractions, injuries and most surgical procedures.

Written by:

Brenda Riske, MS, MBA, MPA Regional Coordinator Mountain States Regional Hemophilia & Thrombosis Center University of Colorado at Denver and Health Sciences Center

Nick Sandmeyer, RN **Nurse Coordinator** Idaho Regional Hemophilia Center

Maribel Johnson, RN, MA **Puget Sound Blood Center** Hemophilia Program

Angela Stewart, RN Nurse Coordinator Alaska Hemophilia Association & Treatment Center

Shirley J. Bleak, RN, FNP Primary Children's Medical Center

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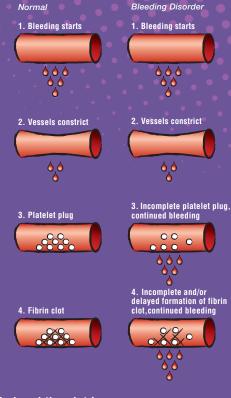
Q: How does the body usually stop bleeding?

A: First, let's talk about normal coagulation (a complex process by which blood forms solid clots)! Take a look at the chart below. This chart shows how clotting occurs normally.

Clot makers vs. Clot breakers

When an injury occurs or a surgical procedure is performed, these things need to happen to stop bleeding and keep it stopped:

- Blood vessels constrict or shrink.
 This slows down the flow of blood through the injured blood vessel (vein or artery).
- 2. Platelets are drawn to the area of injury, become "sticky," and temporarily "plug-up" the injured area of the blood vessel.
- 3. Clotting factors (Clot Makers) interact to form a FIRM fibrin clot, which is like a "scab".
- 4. Fibrinolysis (Clot Breakers), the process in which a fibrin clot is broken down, starts immediately to keep the clot from growing and causing problems. Healing involves both clot making and clot breaking. These occur at the same time.
- 5. The blood vessel starts to heal under the clot. As it heals, the clot gets smaller and smaller until the cut is healed and the clot is gone.



Hemophilia makes it hard for the clot makers to keep up with the clot breakers. This is how "delayed" and /or prolonged bleeding occurs.









Q: Is all hemophilia the same?

A: No. When one has hemophilia, one of the important clot makers, called **clotting factors**, is missing from the blood. The most common clotting factors are Factor VIII (8) and Factor IX (9). People with low levels of Factor VIII have **hemophilia A**. People with low levels of Factor IX have **hemophilia B**. Regardless of what type of hemophilia one has, hemophilia bleeding symptoms can be mild, moderate, or severe, depending on the amount or level of clotting factor produced in their blood.

Q: Is hemophilia contagious?

A: No. It is something that a person has from birth. Consider the following example.

A person's body has genes that are inherited from their mom and dad. Genes are like recipes in a cookbook.

There are gene recipes that determine a person's blood

type, how tall a person is, a person's hair color, and if a person has mild hemophilia. Genes are carried on structures inside cells called chromosomes. Females have 2 X chromosomes (XX) and males have 1 X chromosome and 1 Y chromosome (XY). The gene or "recipe" for mild hemophilia is carried on the X chromosome.

A mom who carries the mild hemophilia gene has a 50/50 chance of passing the gene on to her daughters or sons.

Possible Children of Carrier Mom

(XX-Hemophilia gene is on red X chromosome.)

XX (Mom) + **XY** (Dad) =

Daughters: XX or XX (Carrier daughter = XX; non-carrier daughter = XX) Sons: XY or XY (Son with hemophilia = XY); son without hemophilia = XY)

A father with mild hemophilia will pass the gene to ALL of his daughters. He cannot pass hemophilia on to his sons!

Possible Children of Dad with Hemophilia

XX (Mom) + XY (Dad) =

Daughters: **XX** or **XX** (Carrier daughter = **XX**) Sons: **XY** or **XY** (No sons with hemophilia)

Most often hemophilia is inherited from a parent, but it is possible for hemophilia to just show up with no family history. This is called a "spontaneous mutation."

Q: Are there treatments for mild hemophilia?

A: Yes. There are several medications available to treat bleeding episodes in mild hemophilia.

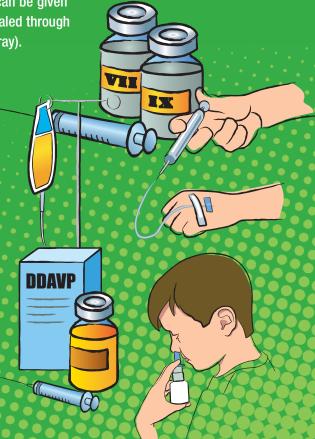
Some medications work by increasing the amount of clotting factor (clot makers) in the blood.

1. Factor VIII or Factor IX can be given into a vein to make the level of blood clotting factor normal until the bleeding stops.

2. DDAVP is a hormone that provides a temporary boost in Factor VIII in the blood. This treatment is only used for people with Hemophilia A. DDAVP can be given into a vein (injection) or inhaled through the nose (Stimate Nasal Spray).

There are also medications that work by stopping the clot breakers.

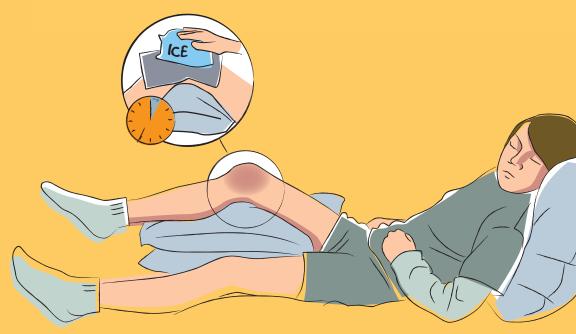
1. Amino caproic acid can be swallowed or given into a vein, however the body has to make a clot before this medication can keep it from breaking down. Thus, either clotting factor or DDAVP must be taken first.



Q: What should a person do if they get injured?

A: There are several treatments available.

- FIRST, call a Treatment Center to discuss whether to use one of the medications listed above.
- SECOND, apply R.I.C.E. first aid as soon as possible to help limit the amount of bleeding and damage. Do this even if clotting factor replacement will also be given.
 - R = REST: The arm or leg should rest on pillows or be put in a sling or bandage. The person should not move the bleeding limb or walk on it.
 - I = ICE: Wrap an ice pack in a damp towel and put it over the bleed. After 5 minutes, remove the ice for at least 10 minutes. Keep alternating: 5 minutes on, 10 minutes off, for as long as the joint or muscle feels hot. This may help decrease pain and limit the amount of bleeding.
 - C = COMPRESSION: The injured area can be wrapped in a tensor bandage "Ace wrap" or elastic stocking to limit bleeding and provide support. Use compression carefully with muscle bleeds if a nerve injury is suspected.
 - **E** = **ELEVATION:** Raise the area that is bleeding above the level of the heart to slow blood loss and limit the amount of swelling.



Q: What are some common myths about mild hemophilia?

1. If I get hurt, I will bleed to death!!!!!

FACT – A person with hemophilia does not bleed faster than normal, but prolonged bleeding can occur if treatment is not received promptly.

2. If I don't see blood, I don't have to worry!!!!

FACT – Some of the most serious bleeding can occur inside the body. If a person suffers a hard bump to the head, call a Treatment Center for advice IMMEDIATELY.

3. I won't be able to play sports!!!!!

FACT – Exercise is actually good for a person's health. Strong joints and muscles prevent injury and bleeding. Many people with hemophilia swim, bike, ski, snorkel, fish, hike, and play baseball, basketball, soccer, and much, much, more!

Q: What are the keys to staying healthy and feeling GOOD?

Visit a Hemophilia Treatment Center once a year.

Brush, floss and see a dentist twice a year.

Keep immunizations up-to-date.

- Stay active! A strong body can avoid injury AND bleeding.
- Eat a variety of foods to maintain a healthy weight and body.
- Avoid risky behaviors. Everyone gets ONE body so take care of it!
- And remember... if an injury does occur, call a treatment center.

Q: Where can one learn more about mild hemophilia?

Research is ongoing into the treatment of mild hemophilia. Visit a hemophilia treatment center (HTC) at least once a year to learn about current recommendations for treating different types of hemophilia.

Here are some educational resources:
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NHF website: www.hemophilia.org
CDC website: www.cdc.gov





NATIONAL HEMOPHILIA FOUNDATION

for all bleeding and clotting disorders

116 West 32nd St, 11th FI • New York NY 10001 www.hemophilia.org • meetings@hemophilia.org

The National Hemophilia Foundation is dedicated to finding better treatments and cures for bleeding and clotting disorders and to preventing the complications of these disorders through education, advocacy and research. Its programs and initiatives are made possible through the generosity of individuals, corporations and foundations as well as through a cooperative agreement with the Centers for Disease Control and Prevention (CDC).