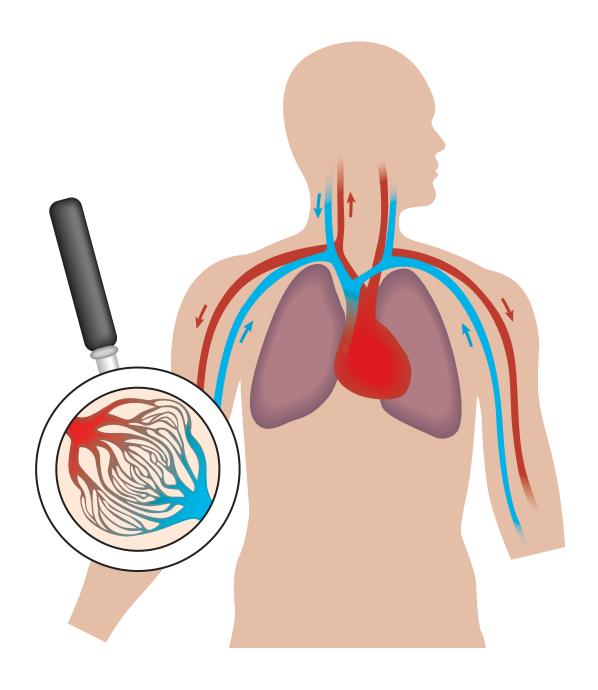


Hemophilia in Pictures





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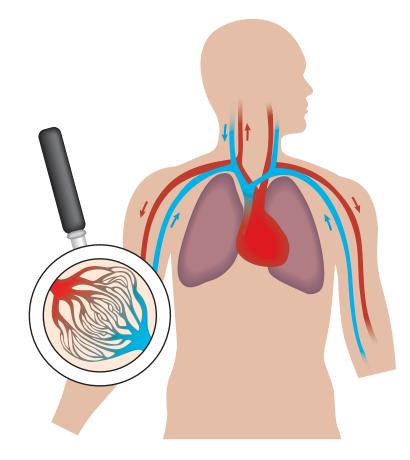
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Hemophilia in Pictures

Learn basic information about hemophilia (an inherited bleeding disorder) using pictures and easy-to-read text.

This publication was developed for people with hemophilia, their caregivers, and healthcare professionals.

Published by the World Federation of Hemophilia



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This publication was developed and tested in large part by volunteers from around the world, most of whom are experts in hemophilia through profession or experience as a person living with the disorder. Volunteers from over 75 hemophilia organizations and treatment centres responded to our survey about Hemophilia in Pictures (HIP). More volunteers tested the new web program and provided feedback on readability and the "user-friendliness" of the program. The WFH Nurses Committee and the Musculoskeletal Committee generously committed time to review the 1998 edition and make recommendations for the new version.

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Table of Contents

PART 1: INTRODUCTION TO HEMOPHILIA

- 1. How does blood move around the body?
- 2. How does bleeding start and stop?
- 3. Why do people with hemophilia sometimes bleed longer than other people?
- 4. Is all hemophilia the same?
- 5. How do people get hemophilia?
- 6. What are the chances that a baby will have hemophilia?
- 7. Is hemophilia lifelong?

PART 2: ASSESSING AND MANAGING BLEEDS

- 8. What are some common signs of hemophilia?
- 9. What causes a joint bleed?
- 10. What happens in a joint bleed?
- 11. Which joint bleeds are most common?
- 12. What are the long-term effects of joint bleeds?
- 13. What causes a muscle bleed?
- 14. What happens in a muscle bleed?
- 15. Which muscle bleeds are most common?
- 16. What are the long-term effects of muscle bleeds?
- 17. Which bleeds are serious or life-threatening?

PART 3: TREATMENT OF BLEEDS

- 18. Why should bleeds be treated quickly? (part A)
- 19. Why should bleeds be treated quickly? (part B)
- 20. How can bleeds be treated with first aid?
- 21. How can bleeds be treated with factor replacement therapy?
- 22. What other treatment may help?
- 23. What are inhibitors and how can they be treated?
- 24. What are the signs of recovery from a bleed?

PART 4: STAYING HEALTHY AND PREVENTING BLEEDS

- 25. What can be done to stay healthy?
- 26. Why is dental health important?
- 27. Why is emotional health important?
- 28. What if an operation is needed?
- 29. What if medicine or vaccinations are needed?
- 30. What are some allergic reactions to treatment?
- 31. What medical information should be carried?
- 32. Where can people get help or advice about hemophilia?

ANNEX: HOME THERAPY AND VENEPUNCTURE

- 33. What is home therapy?
- 34. What is venepuncture?
- 35. What preparation is needed for replacement therapy?
- 36. How can infection be prevented when giving an injection?
- 37. How is replacement therapy given? (part A)
- 38. How is replacement therapy given? (part B)

ADDITIONAL RESOURCES

Glossary



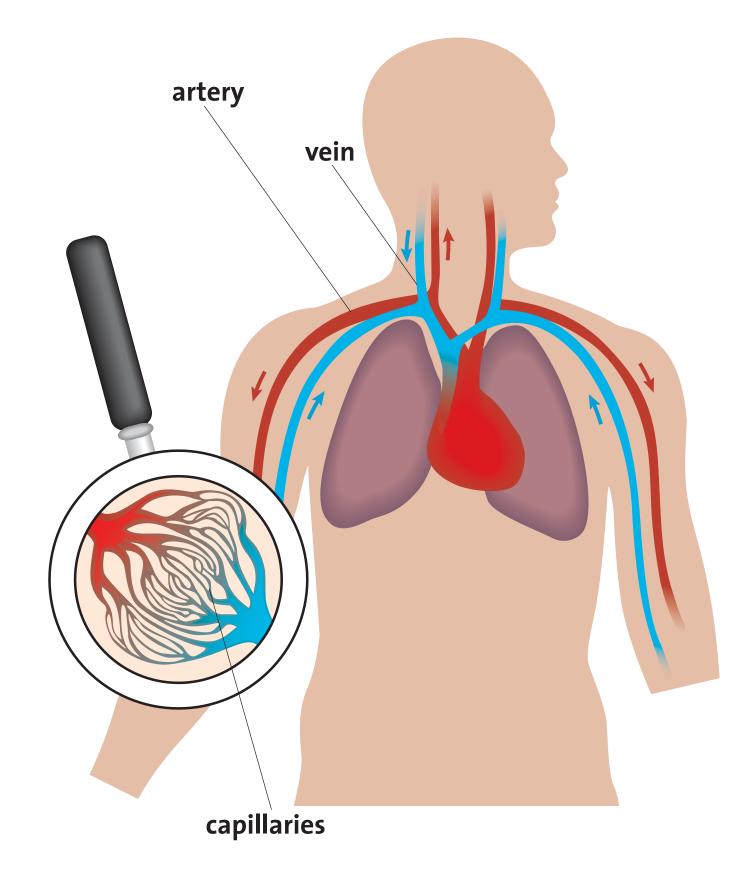
Part 1

Introduction to Hemophilia



How does blood move around the body?

- Hemophilia is a bleeding disorder, so it is important to understand the blood system.
- The heart pumps blood around the body.
- Blood moves through the body in tubes called arteries, veins, and capillaries. Some are large (arteries and veins) and some are small (capillaries).

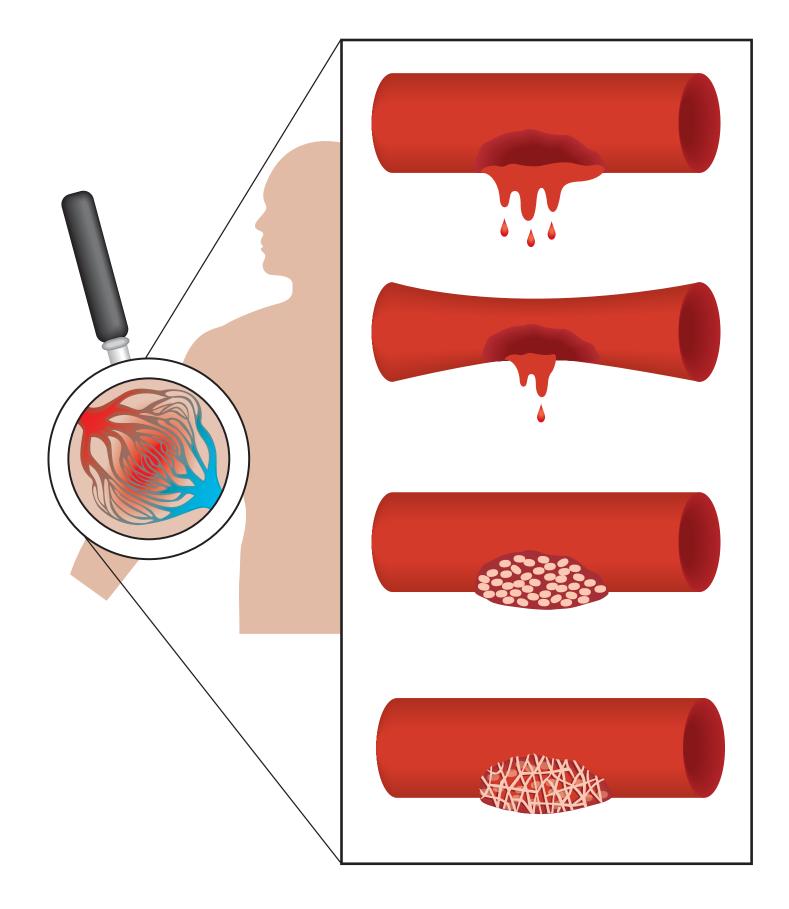




1.

How does bleeding start and stop?

- Bleeding starts when a capillary is injured and blood leaks out.
- The capillary tightens up to help slow the bleeding.
- Then blood cells called *platelets* make a plug to patch the hole.
- Next, many *clotting factors* in *plasma* (part of the blood) work together to form a clot over the plug. This makes the plug stronger and stops the bleeding.



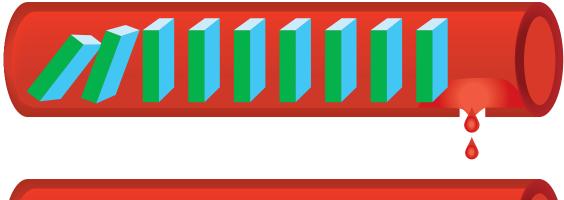


Why do people with hemophilia sometimes bleed longer than other people?

- In hemophilia, one clotting factor is missing, or the level of that factor is low. This makes it difficult for the blood to form a clot, so bleeding continues longer than usual, not faster.
- Since there are many clotting factors in plasma, each factor is named with a Roman numeral.

Example:

VIII = eight IX = nine

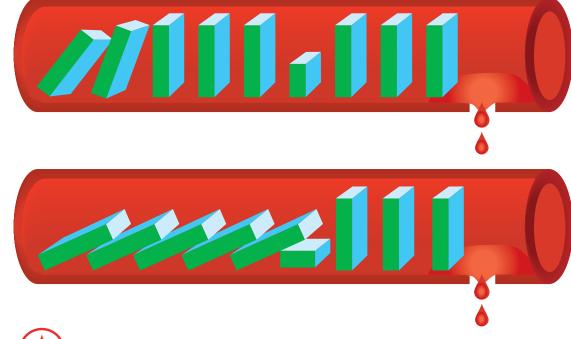


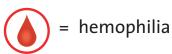


Clotting in hemophilia

Normal clotting process







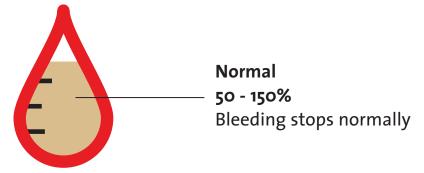




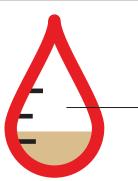
Is all hemophilia the same?

- People with low levels of factor VIII (eight) have *hemophilia A*.
- People with low levels of factor IX (nine) have *hemophilia B*.
- Hemophilia can be mild, moderate, or severe, depending on the level of clotting factor.

Degrees of Severity







Mild Hemophilia 5 - 40%

If factor level is over 40%, bleeding usually does not continue longer than normal.

If factor level is under 40%, bleeding may continue longer than normal, usually after an injury or surgery.



Moderate Hemophilia

1 - 5%

Bleeding will continue longer than normal after an injury or surgery.

Bruising or bleeding can happen easily.



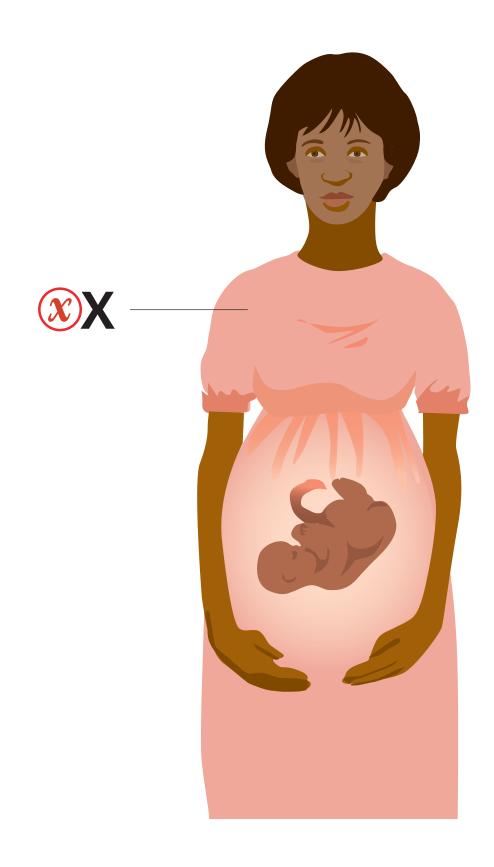
Severe Hemophilia usually less than 1%

Bleeding can happen for no clear reason.



How do people get hemophilia?

- People are born with hemophilia. They **cannot** catch it from someone.
- Hemophilia is usually inherited, meaning that it is passed on through a parent's *genes*. Genes carry messages about the way the *cells* of the body work. For example, they determine a person's hair and eye colour.

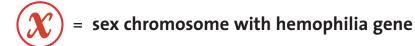


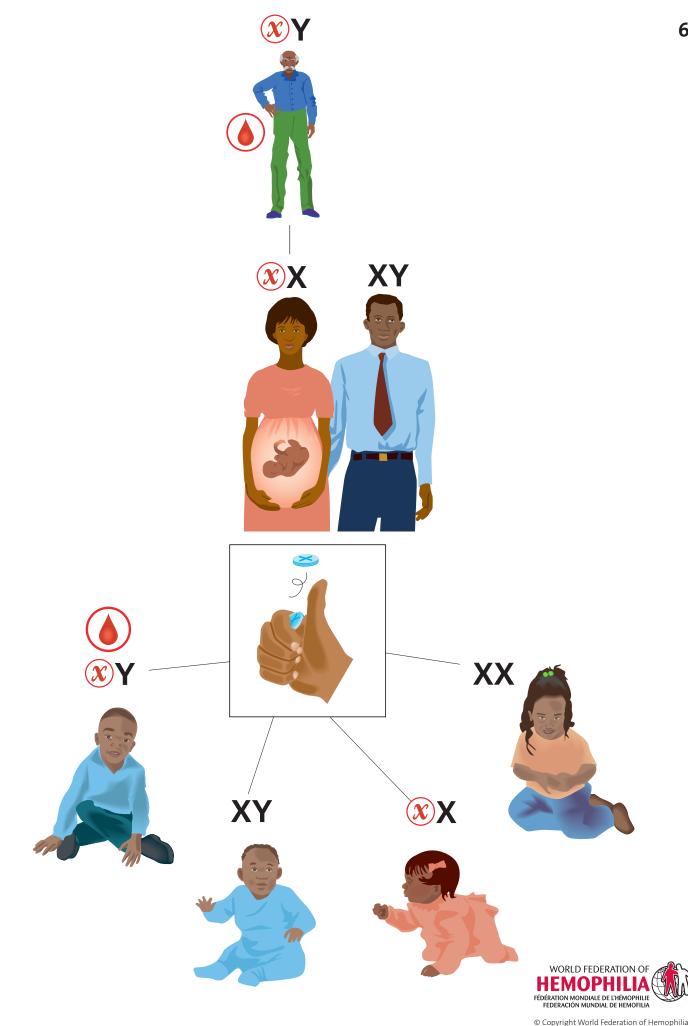




What are the chances a baby will have hemophilia?

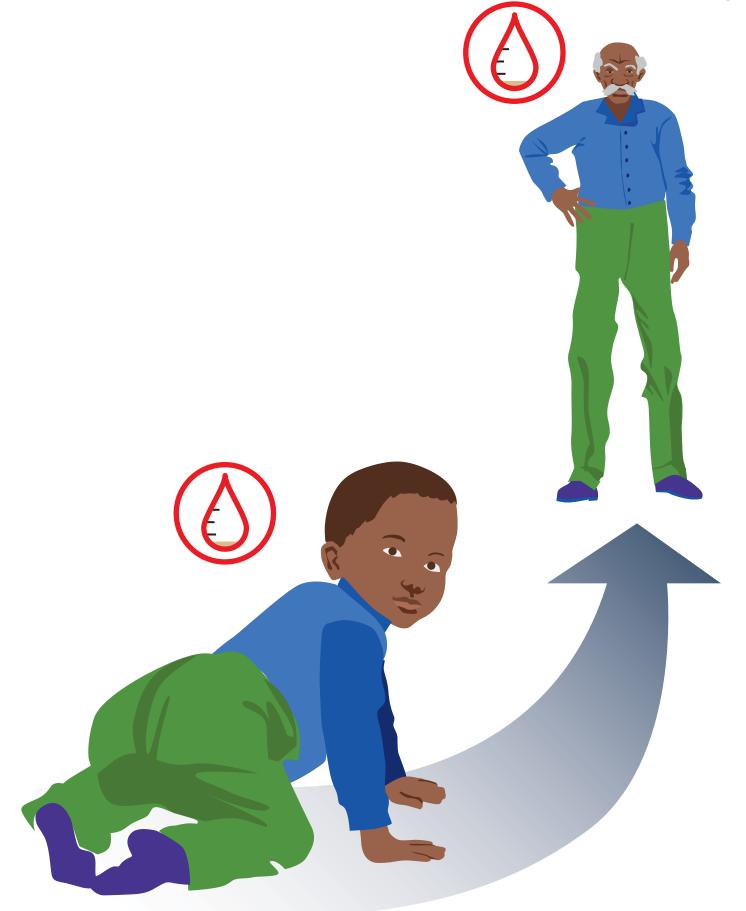
- Genes are found on *chromosomes*. Two of these chromosomes (called X and Y) decide a person's sex. Females are born with two Xs. Males are born with one X and one Y.
- The hemophilia gene is carried on the X chromosome.
- A man with hemophilia passes the hemophilia gene to all of his daughters, but not to his sons. His daughters are called *carriers* because they carry the hemophilia gene.
- When a carrier has a baby, there is a one in two chance that she will pass on the hemophilia gene. If she passes the hemophilia gene to a son, he will have hemophilia. If she passes the hemophilia gene to a daughter, she will be a carrier like her mother.
- Sometimes a baby is born with hemophilia although his mother is not a carrier. This is because the factor VIII or IX gene changed only in the baby's body. One in three babies born with hemophilia have no family history of it.





Is hemophilia lifelong?

- A person born with hemophilia will have it for life.
- The level of factor VIII or IX in his blood usually stays the same throughout his life.







Part 2

Assessing and Managing Bleeds

What are some common signs of hemophilia?

- In hemophilia, bleeding can happen anywhere in the body. Sometimes it can be seen and sometimes it cannot.
- Bleeding can happen after an injury or surgery. It can also happen for no clear reason. This is called *spontaneous bleeding*.
- Bleeding is rare in babies with hemophilia, but they may bleed for a long time after circumcision.
- When babies start to walk, they bruise easily. They also bleed longer than usual after an injury, especially to the mouth and tongue.
- As children grow, spontaneous bleeding becomes more common. It affects the joints and muscles.



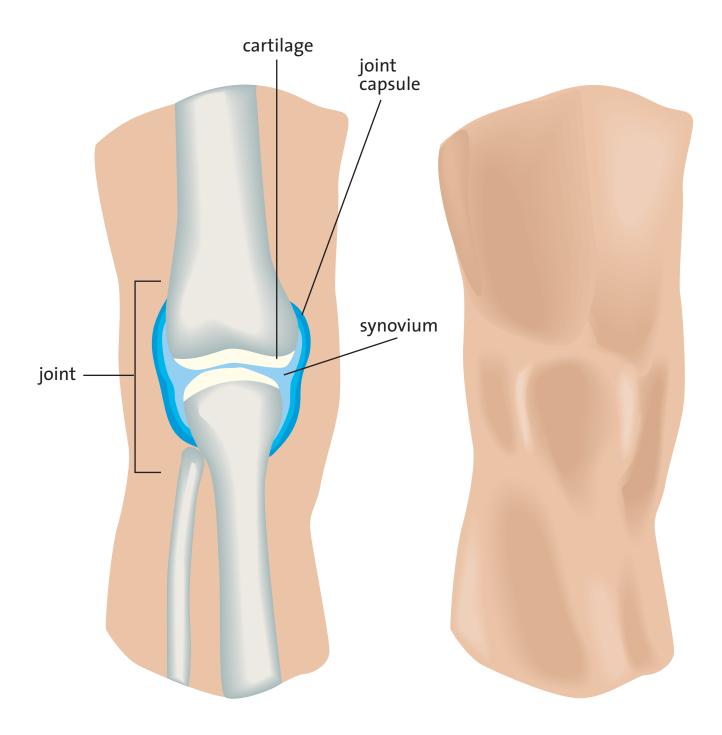




What causes a joint bleed?

- The place where two bones meet is called a *joint*. The ends of the bones are covered with a smooth surface called *cartilage*.
- The bones are partly held together by a *joint capsule*. The joint capsule has a lining called *synovium* with many capillaries (small blood vessels). It makes a slippery, oily fluid that helps the joint move easily.
- If the capillaries in the synovium are injured, they bleed.

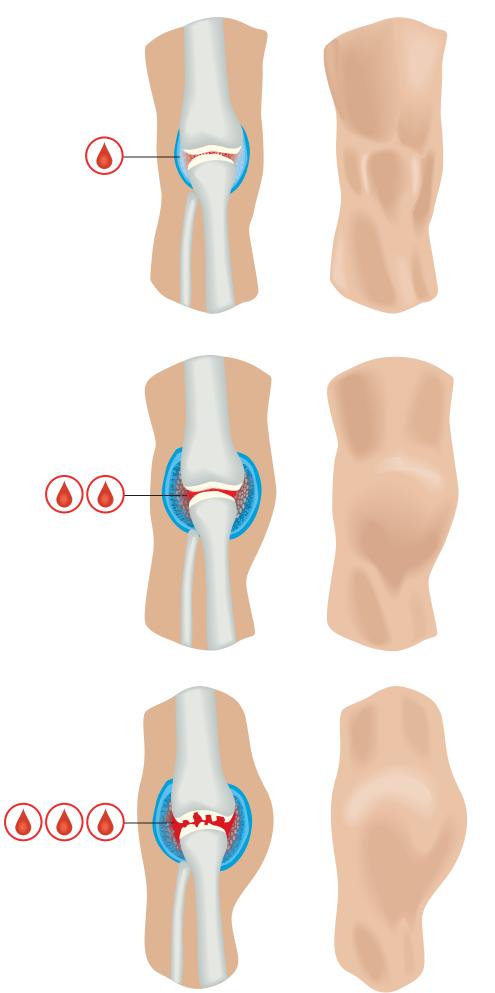
 Often there is no clear reason for the bleed, especially in severe hemophilia. In a person who does not have hemophilia, the clotting system stops the bleeding quickly. But in hemophilia, the bleeding continues. This causes the joint to swell and become painful.





What happens in a joint bleed?

- A person with hemophilia knows when a bleed starts because the joint feels tingly and warm.
- As blood fills the capsule, the joint swells and becomes painful and hard to move.
- Without treatment, the pressure from the swelling eventually stops the bleed. Later, special cells clear most of the blood out of the joint.



Bubbling Tingling Heat

Swelling Pain Heat

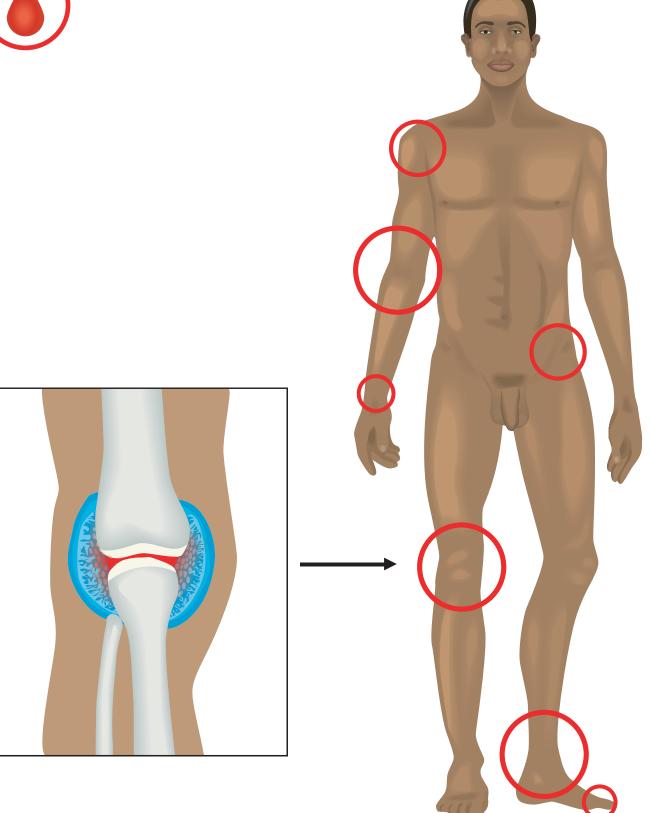
Boggy Swollen Muscle Wasting Morning Stiffness Chronic Pain Limited Movement



Which joint bleeds are most common?

- The most common joint bleeds happen in ankles, knees, and elbows.
- Bleeds into other joints can also happen, including the toes, shoulders, and hips.
- Joints of the hands are not usually affected except after injury.

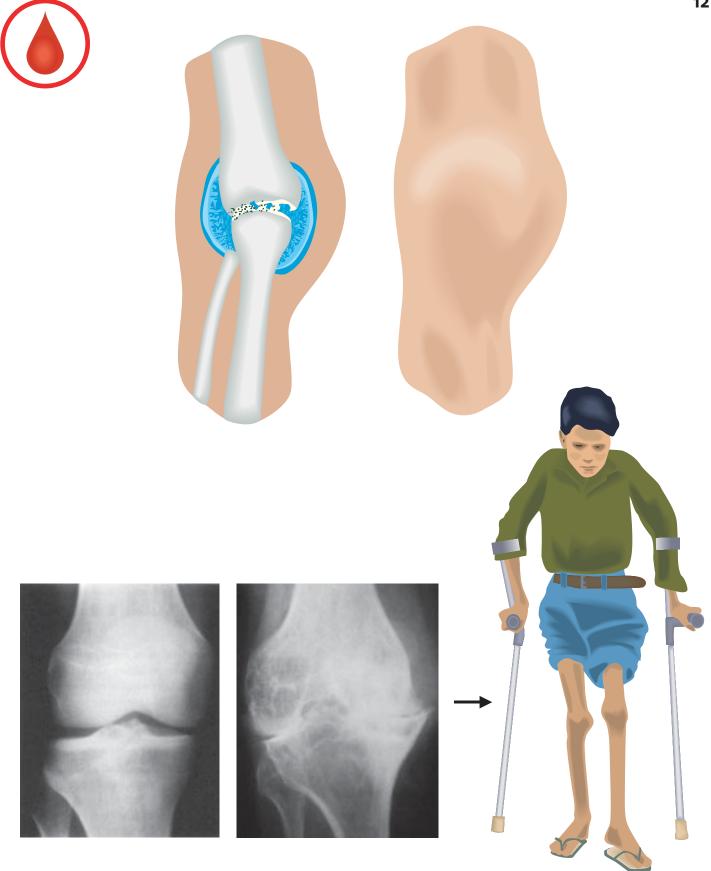






What are the long-term effects of joint bleeds?

- Repeated bleeding into a joint causes the synovium (lining) to swell and bleed very easily.
- Some blood remains in the joint after each bleed. The synovium stops producing the slippery, oily fluid that helps the joint move.
- This damages the smooth cartilage that covers the ends of the bones. The joint becomes stiff, painful to move, and unstable. It becomes more unstable as muscles around the joint weaken.
- With time, most of the cartilage breaks down and some bone wears away. Sometimes the joint cannot move at all. The whole process is called *hemophilic arthritis*.





What causes a muscle bleed?

- Muscle bleeds happen when capillaries in the muscle are injured.
- Sometimes the cause is known, but bleeds can also happen for no clear reason.

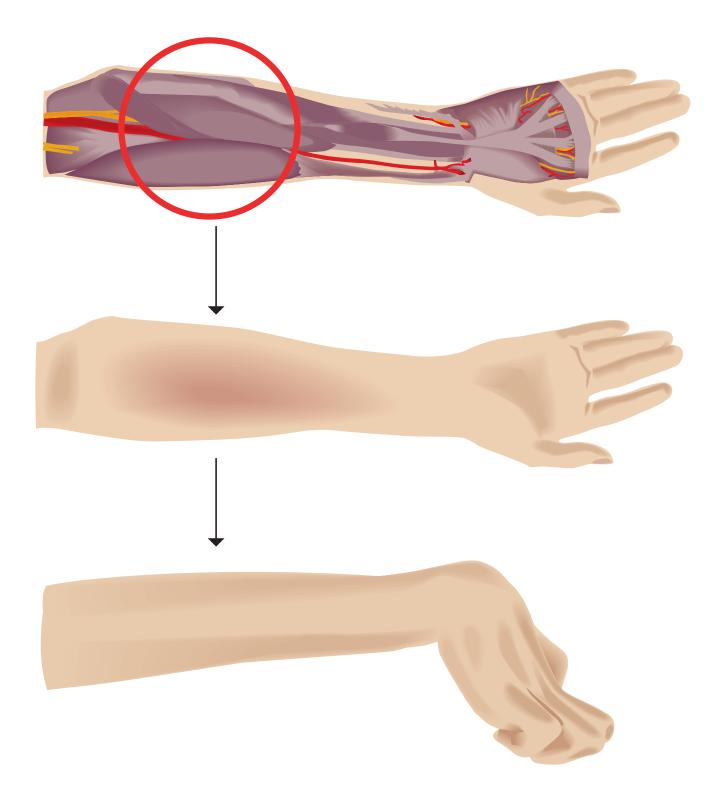




What happens in a muscle bleed?

- During a bleed, the muscle feels stiff and painful.
- The bleed causes swelling that is warm and painful to touch. There may be bruising if the bleed is near the skin.
- In some of the deeper muscles, the swelling may press on *nerves* or *arteries*, causing tingling and numbness.
- The muscle tightens up to protect itself. This is called a *muscle spasm*. As a result, joints that are usually moved by that muscle do not move properly.



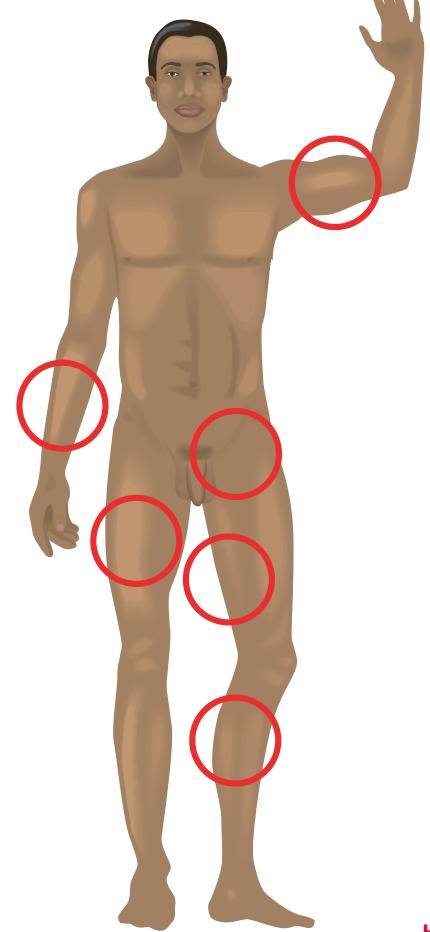




Which muscle bleeds are most common?

- Muscle bleeds happen in the *calf, thigh,* and *upper arm.*
- Bleeds in the psoas muscle (at the front of the hip) and the forearm muscles are also common. These bleeds can put pressure on nerves and arteries, causing permanent damage.
- Bleeds into the muscles of the hands are rare and usually follow an injury.

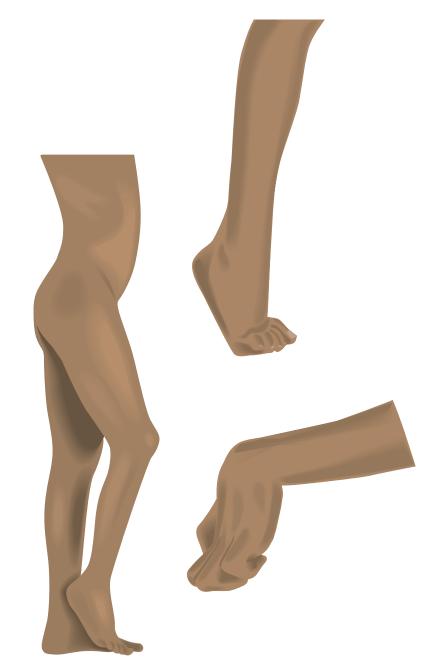


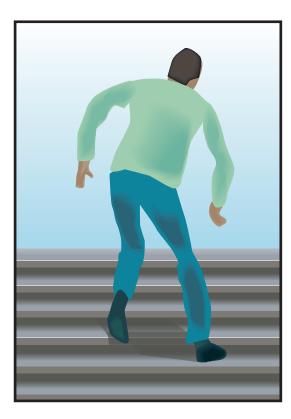


What are the long-term effects of muscle bleeds?

- After repeated bleeds, muscles can become weak, scarred, and shorter than normal (sometimes permanently). They can no longer protect the joints.
- Joints above and below the muscle cannot move properly. They may bleed more often.
- If nerves are damaged during muscle bleeds, the muscle may become weak or even paralyzed.
- Permanent damage to joints, muscles, and nerves affects the way a person sits, stands, and walks.



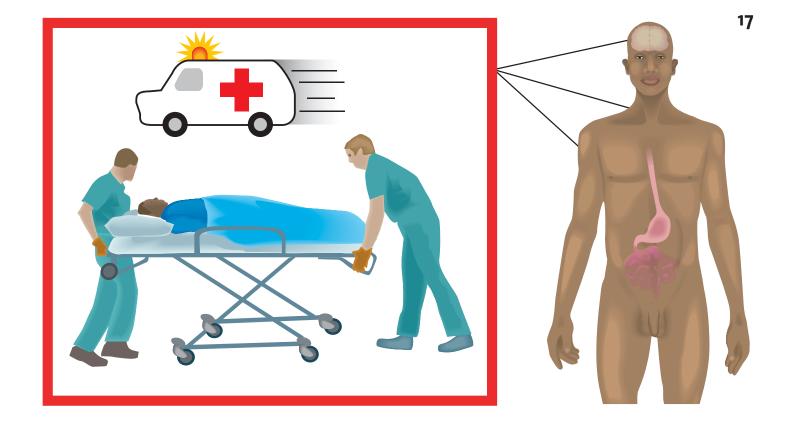


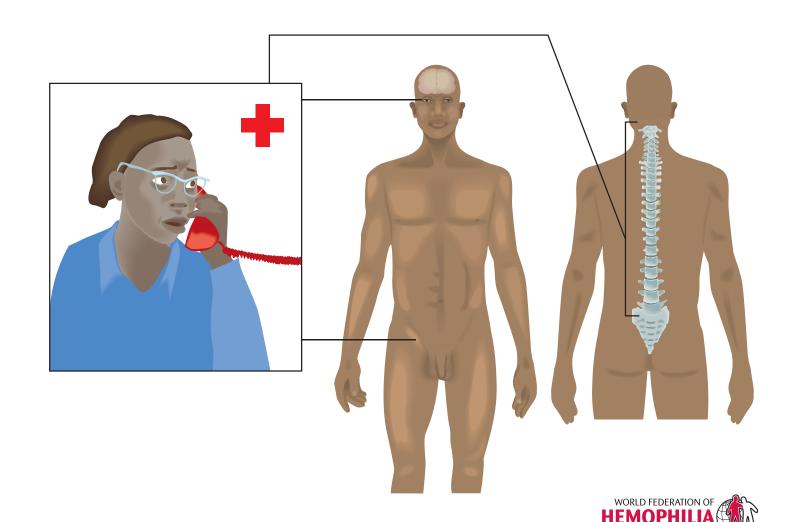




Which bleeds are serious or life-threatening?

- Bleeding within the head (usually resulting from injury) is a major cause of death in hemophilia, especially in children. Head bleeds can cause headache, nausea, vomiting, sleepiness, confusion, clumsiness, weakness, fits, and loss of consciousness.
- Bleeding into the throat may result from infection, injury, dental injections, or surgery. Throat bleeds cause swelling, as well as difficulty swallowing and breathing.
- Major loss of blood is life-threatening. It is uncommon in hemophilia except after an injury or when related to another medical condition.
- Other bleeds may be very serious, but usually not lifethreatening, such as bleeds into the eyes, spine, and psoas muscle.
- Blood in the urine is common in severe hemophilia, but rarely dangerous.





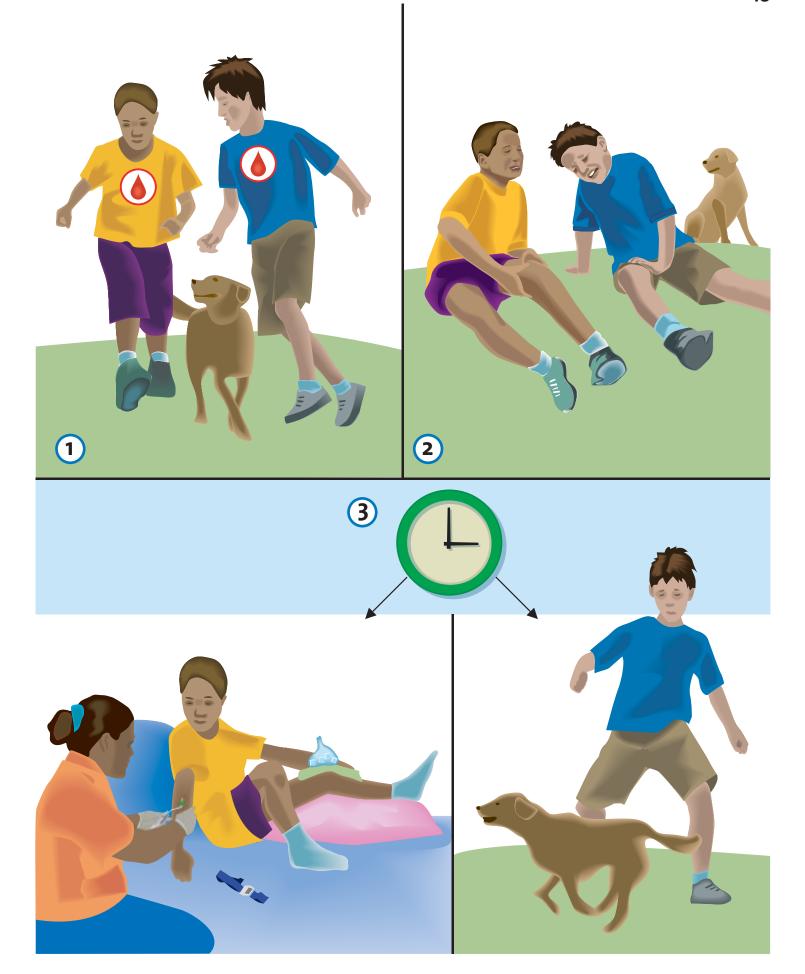


Part 3

Treatment of Bleeds

Why should bleeds be treated quickly? (PART A)

- Bleeds should be treated quickly to recover more quickly and prevent later damage.
- If in doubt, treat. Don't wait!



Why should bleeds be treated quickly? (PART B)

■ When treatment is late, the bleed takes longer to heal, and more treatment product is needed.



How can bleeds be treated with first aid?

Start first aid as soon as possible to limit the amount of bleeding and damage. Do this even if factor replacement will also be given.

REST:

The arm or leg should rest on pillows or be put in a sling or bandage. The person should not move the bleeding joint or walk on it.

■ ICE:

Wrap an ice pack in a damp towel and put it over the bleed. After five minutes, remove the ice for at least ten minutes. Keep alternating: five minutes on, ten minutes off, for as long as the joint feels hot. This may help decrease pain and limit bleeding.

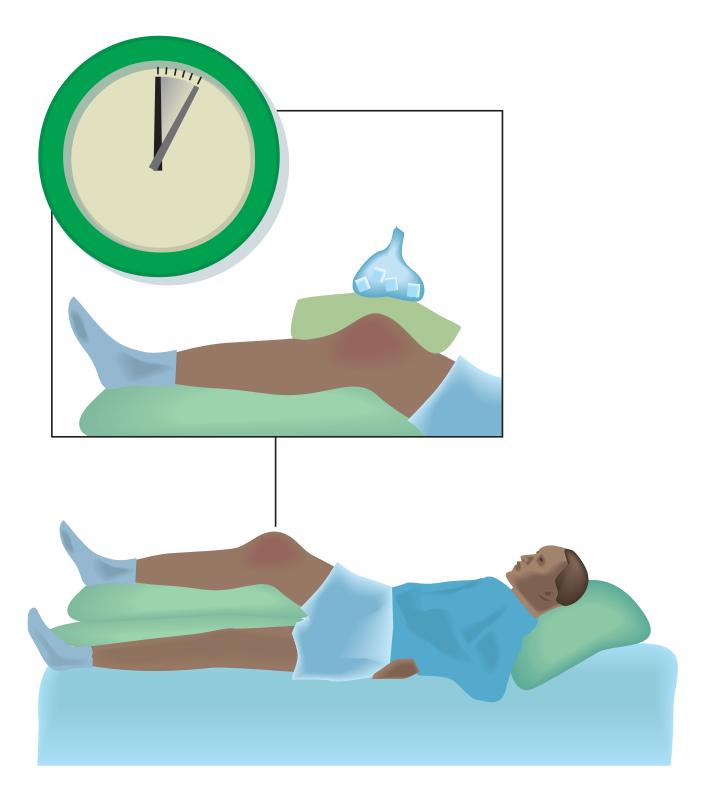
COMPRESSION:

Joints can be wrapped in a tensor bandage or elastic stocking. This gentle pressure may help to limit bleeding and support the joint. Use compression carefully with muscle bleeds if a nerve injury is suspected.

■ ELEVATION:

Raise the area that is bleeding above the level of the heart. This may slow blood loss by lowering pressure in the area.

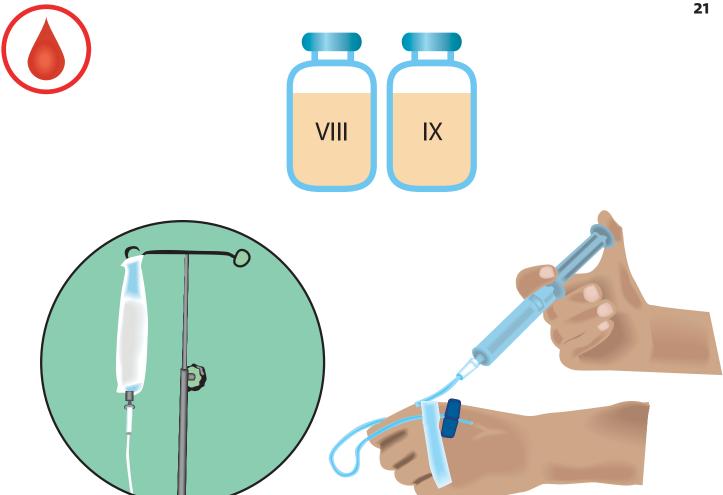


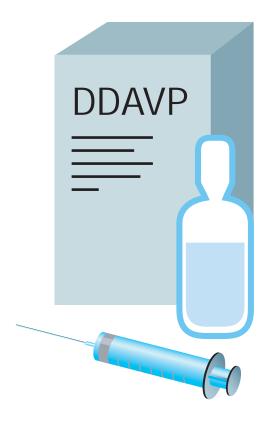




How can bleeds be treated with factor replacement therapy?

- Hemophilia can usually be treated by injecting the missing clotting factor into a vein. Clotting factor cannot be given by mouth.
- Clotting factor can be found in various treatment products, such as *cryoprecipitate* and *factor concentrate*. The possible side effects of a product should always be considered before using it.
- People with mild hemophilia A (or another condition called von Willebrand disease) can be treated with a medicine called desmopressin or DDAVP. It can be given by injection into a vein, injection under the skin, or nasal spray.
- Repeated treatment is usually needed.



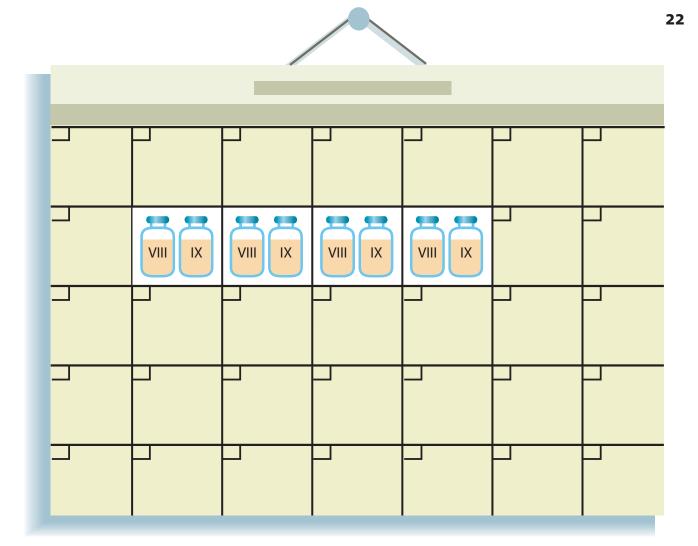






What other treatment may help?

- Other treatments may help, such as:
 - Pain medication;
 - Anti-inflammatory medication to reduce swelling;
 - A different dose or schedule of factor replacement; and
 - Repeated doses of factor VIII or IX.
- A physiotherapist can:
 - Suggest ways to strengthen muscles and restore joint movement;
 - Say if it is safe to return to normal activities; and
 - Suggest ways to prevent further injury.





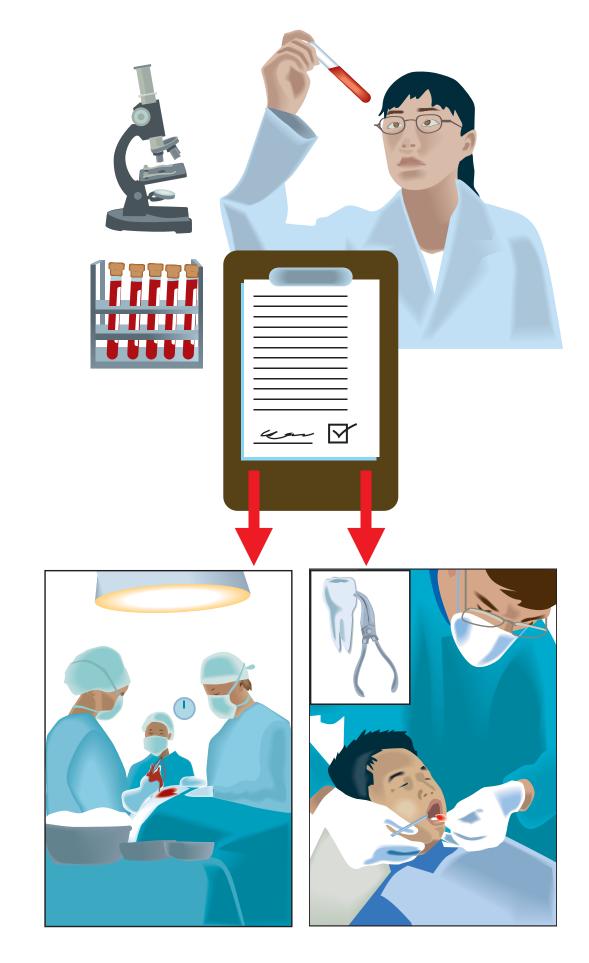




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What are inhibitors and how can they be treated?

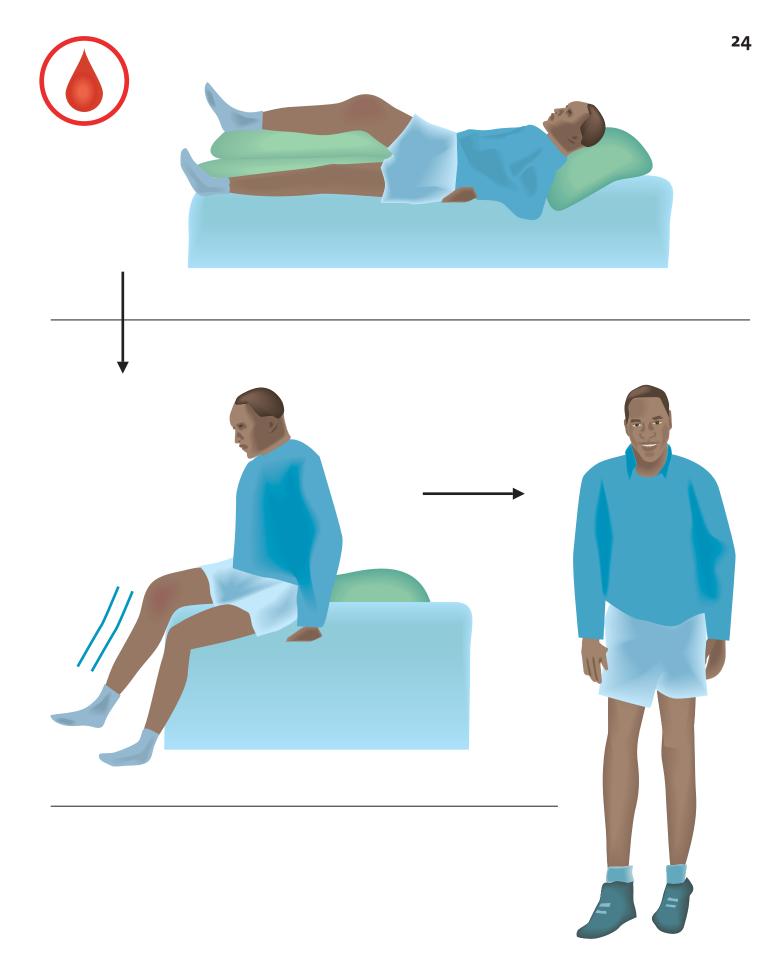
- *Inhibitors* are antibodies (proteins) made by the body to fight off things it sees as "foreign".
- A person with hemophilia may develop inhibitors that fight off the foreign protein in the treatment product. If the inhibitors are strong, the usual treatment may become less effective.
- Inhibitors are not very common. They are most often found in people with severe hemophilia A.
- Test for inhibitors before surgery, including dental surgery.
- Special treatments are available to deal with inhibitors.





What are the signs of recovery from a bleed?

- Full movement of the joint or muscle returns.
- Full strength of the muscle returns.







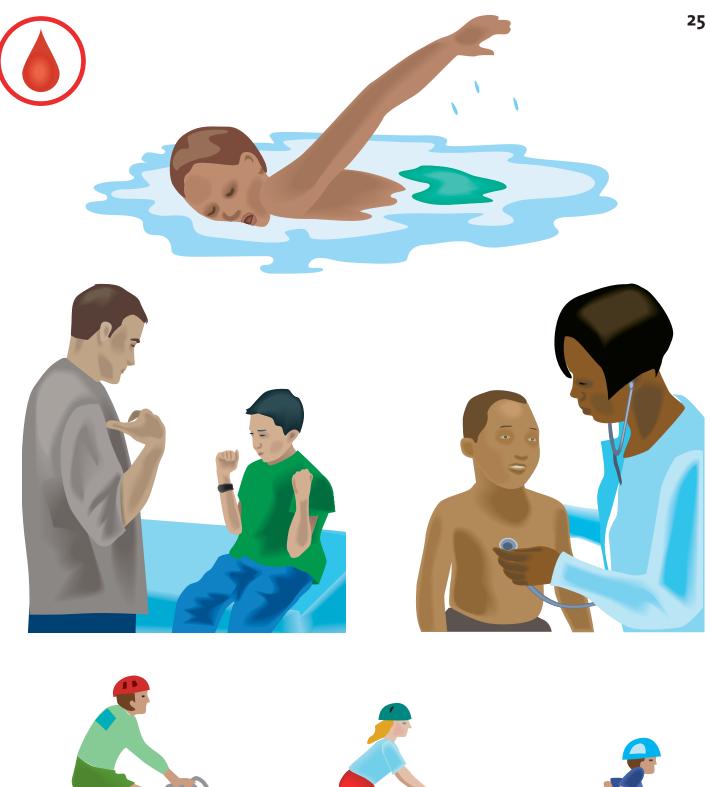
Part 4

Staying Healthy and Preventing Bleeds

What can be done to stay healthy?

Medical treatment is only one part of good health. People with hemophilia should:

- Exercise and stay fit.
- Wear protection that is appropriate for the sport or activity.
- Get regular check-ups that include joint and muscle examination.
- Get all vaccinations recommended, including hepatitis A and hepatitis B protection.
- Maintain a healthy body weight. People who do not exercise are more likely to put on extra weight. A person with hemophilia needs to control his weight so that he does not put extra stress on his joints, especially if he has arthritis.





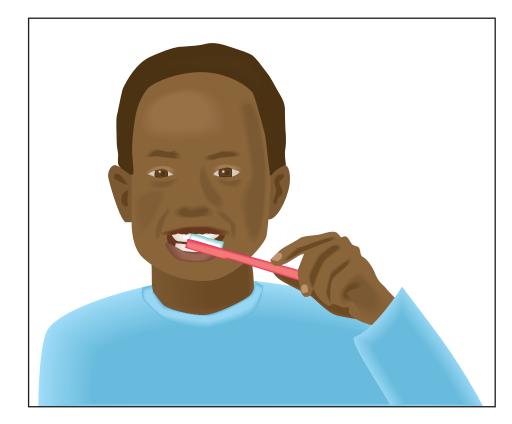






Why is dental health important?

- Healthy teeth and gums reduce the need for hemophilia treatment.
- Regular dental care reduces the need for injections and surgery.
- Dental care should include brushing, flossing, and check-ups by a dentist.







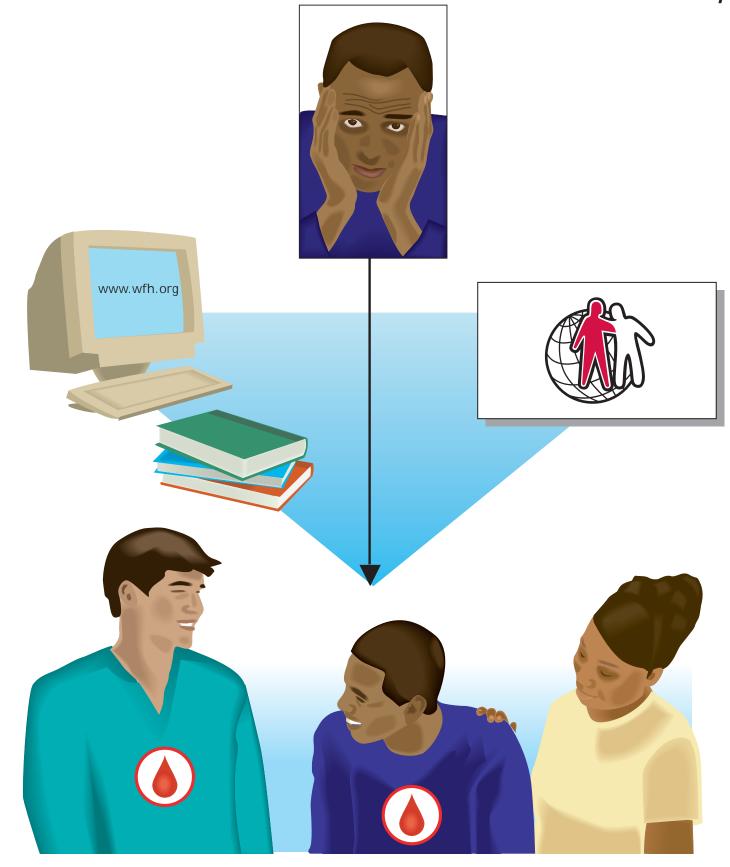


Why is emotional health important?

Living with hemophilia causes stress. This stress can affect family members, as well as the person with hemophilia.

It may help to:

- Learn as much as possible about hemophilia. Knowledge helps people feel more in control.
- Meet other people with hemophilia. Sharing knowledge can reduce stress.
- Join a hemophilia organization.

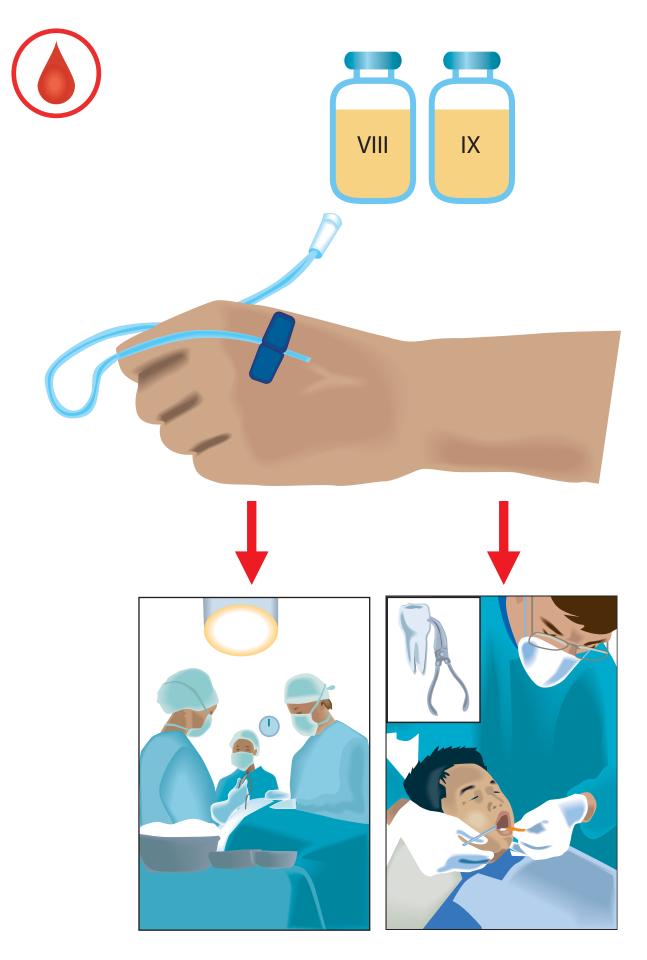




What if an operation is needed?

Before an operation, dental surgery, or dental injections healthcare workers should:

- Obtain enough treatment product to control bleeding for a the procedure, as well as complete recovery.
- Test for inhibitors.
- Consider other medications that may help speed recovery, such as pain killers. Antifibrinolytics can be used to stop the normal breakdown of clots by the body.







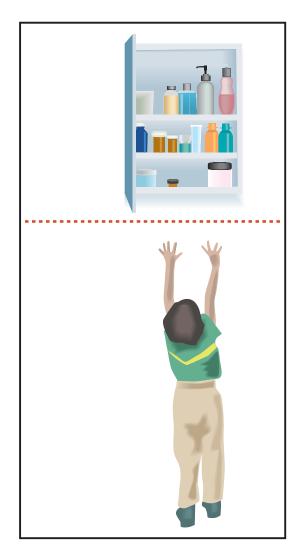
DOs

- Check all medicine with a hemophilia healthcare worker.
- Store and use medicine and treatment products according to instructions.
- Store all medicine out of reach of children.

DON'Ts

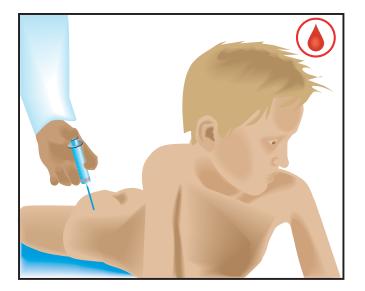
- Do not take ASA (Aspirin®) in any form to reduce pain.
- Do not take *nonsteroidal anti-inflammatory drugs (NSAIDs)* without medical advice.
- Do not get muscle injections. When vaccinations are necessary, they should be given under the skin instead.















What are some allergic reactions to treatment?

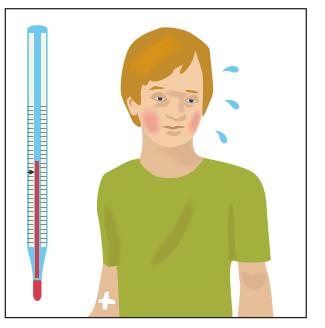
Some treatments can cause an allergic reaction that may result in:

- Fever
- Shivering
- Skin rash

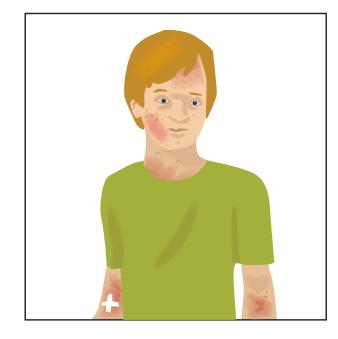
Reactions are usually mild. They can be eased by taking *antihistamines* (medicine that is usually taken as pills).

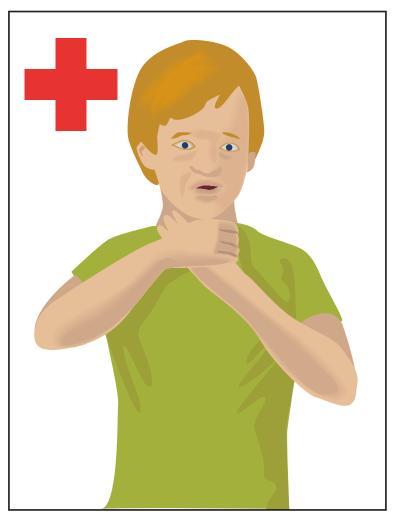
Medical help is needed quickly for:

- Difficulty breathing
- A tight feeling in the chest











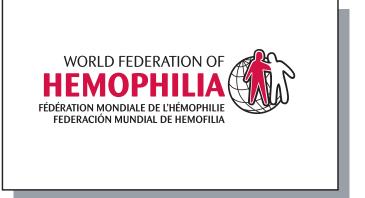


What medical information should be carried?

- A person with hemophilia should carry information about his health, including the type of hemophilia, treatment needed, and allergies.
- An international medical card is available free through the World Federation of Hemophilia. Tags called Medic-Alert and Talisman are sold in some countries.











Where can people get help or advice about hemophilia?

Help and advice is available from:

- Hemophilia healthcare workers
- Hemophilia organizations (listed on the WFH web site)
- Other people with hemophilia and their families (through meetings or telephone help lines)
- Publications about hemophilia







Annex

Home Therapy and Venepuncture

What is home therapy?

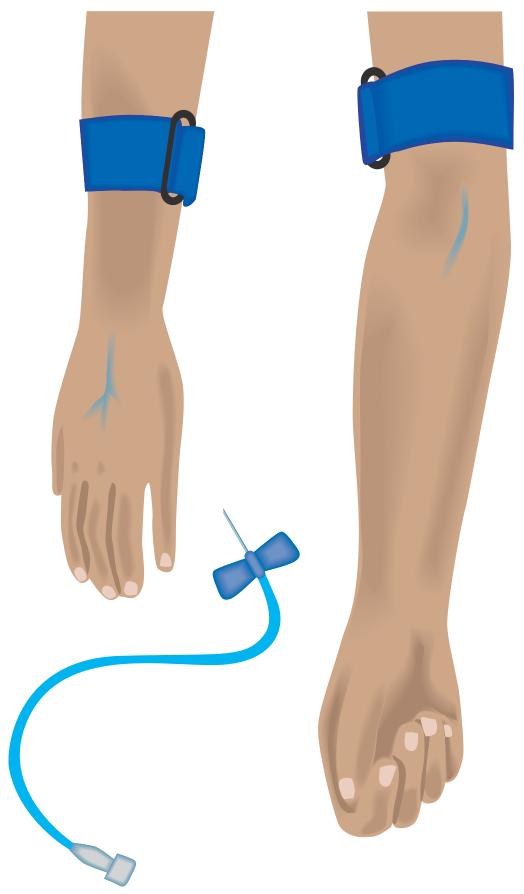
- Home therapy is *infusion* (injection) with clotting factor replacement away from the hospital. A person with hemophilia can infuse at home, school, work, or elsewhere.
- A written record of all treatments must be kept.
- A person with hemophilia and his family share responsibility for their health with the hemophilia healthcare workers.
- Home therapy does not replace a doctor's care.





What is venepuncture?

- *Venepuncture* means to put a needle into a vein. This can be done to take blood, or to give an injection.
- Any vein that can be seen or easily felt can be used for injection. Usually, the easiest veins to use are on the back of the hand or inside the elbow.
- Babies with hemophilia may bleed severely if injected into veins in the neck or groin. These sites are sometimes used by doctors taking blood samples. Other sites should be used in babies with hemophilia, both for samples and for treatment.





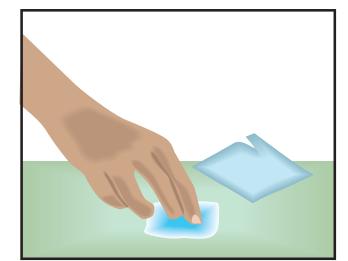
What preparation is needed for replacement therapy?

Read the instructions that came with the factor concentrate. Before handling any materials, wash hands thoroughly with soap and water. The work surface should be cleaned with disinfectant. A capful of chlorine bleach in a half litre of water makes a good disinfectant. Make sure the bleach is not out of date.

Supplies needed for treatment with factor concentrate:

- sharps container
- disposable wipes
- alcohol wipe
- bandage
- cotton balls
- tape
- tourniquet
- butterfly needle

- syringe
- transfer needle/filter needle
- factor concentrate
- latex gloves
- diluent (sterile liquid) supplied with the concentrate



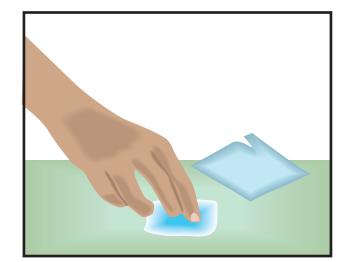




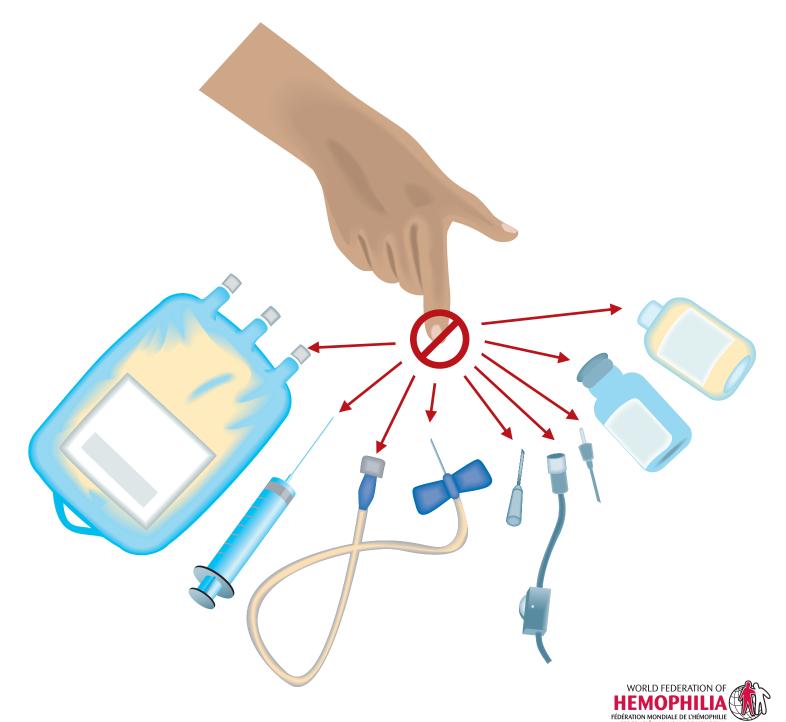
5.

How can infection be prevented when giving an injection?

- Take special care in handling all medical equipment. Keep hands and equipment clean. Helpers should wash hands and wear gloves.
- Be careful! Keep fingers away from ends of needles, all sharp objects, and the openings of bottles, bags, and syringes.
 These areas must be kept very clean.
- Concentrates should not be opened and then stored for later use.
- Place all used needles and syringes into a sharps container.
- Clean any spills with disinfectant, as the hemophilia healthcare worker instructs.
- Safely dispose all materials used during the injection, according to local policy. Check with the closest hemophilia centre or clinic for its recommendations.



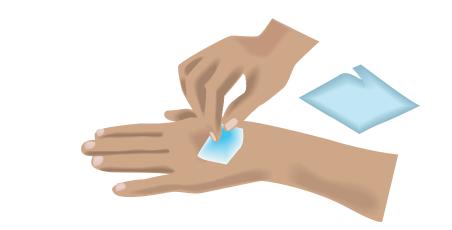




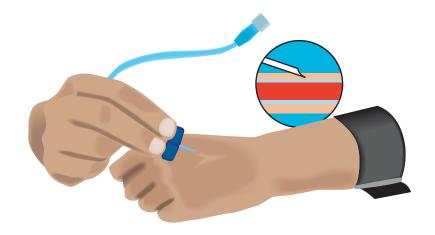
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How is replacement therapy given? (PART A)

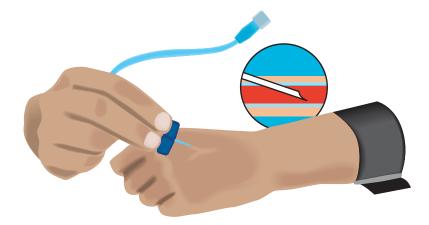
- Be sure that your hands are clean.
- Follow healthcare worker's instructions carefully.
- Use all of the dissolved concentrate. Do not be tempted to split the dose and keep some for later. The clotting factor content begins to disappear after it is reconstituted (after the diluent is added to the concentrate). In addition, there is a high risk of infection.



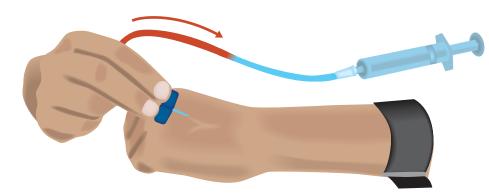
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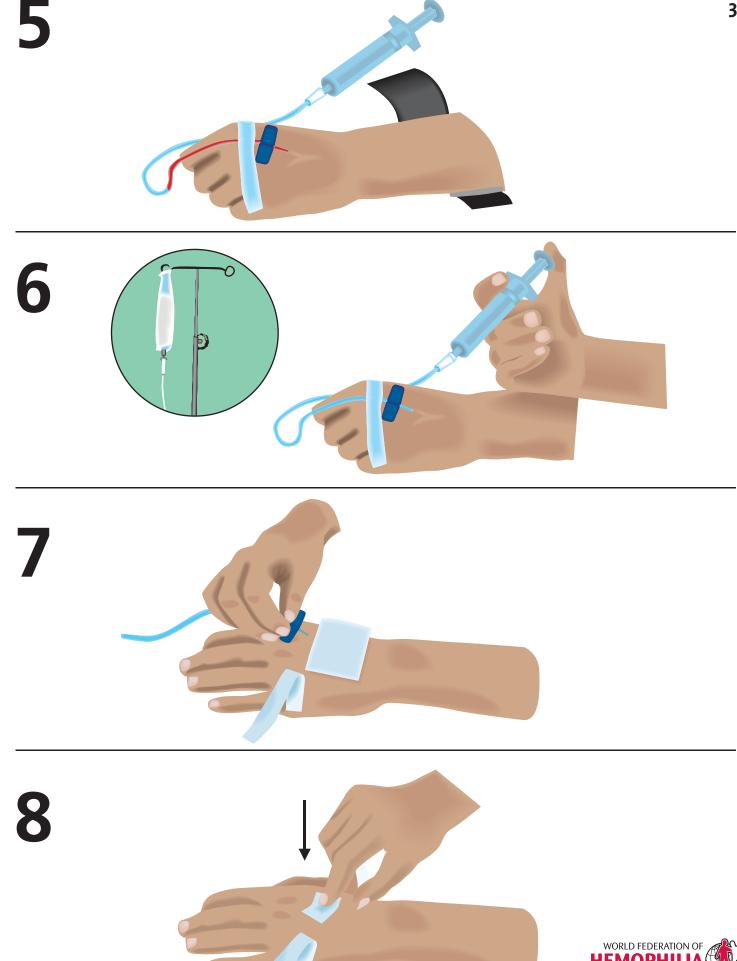


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How is replacement therapy given? (PART B)

- Follow healthcare worker's instructions carefully.
- Remember, once you are finished and the needle is no longer in your vein, press down on the place where the needle went in for at least five minutes.





Glossary

antifibrinolytic, p.28:

A drug that can help stop the normal breakdown of blood clots and help speed recovery from a bleed.

antihistamine, p.30:

A drug that can reduce or prevent allergic reactions.

anti-inflammatory, p.22:

A drug that can reduce or prevent pain, redness, and swelling caused by inflammation (the body's reaction to irritation, injury, or infection).

artery, p.1 & 14:

A large tube or blood vessel that carries blood from the heart through the body. The body has several arteries.

calf, p.15:

The area of the leg between the knee and the ankle.

capillary, p.1:

Any of the very small tubes or blood vessels that form a network to carry blood through the body. The body has many capillaries.

carrier, p.6:

A person who carries a gene that causes a disorder, usually showing no symptoms.

cartilage, p.9:

The smooth surface covering the ends of the bones in a joint.

cell, p.5:

The smallest independent living thing on earth. The human body is made of trillions of tiny cells.

chromosome, p.6:

A very fine, threadlike strand of proteins and DNA in the centre of human, animal, and plant cells. Two chromosomes (called X and Y) decide a person's sex. Females are born with two X chromosomes (XX = girl). Males are born with one X and one Y chromosome (XY = boy).

clot, p.2:

A thick lump of blood formed by clotting factors that work together to help stop bleeding.

clotting factor, p.2:

Any of the factors in blood plasma that work together to form a clot to help stop bleeding.

compression, p.20:

To apply compression to a bleed means to apply firm pressure or support using an elastic stocking or wrap to help stop the swelling.

cryoprecipitate, p.21:

A treatment product made from blood plasma. It contains proteins, such as factor VIII (eight) and von Willebrand factor, but not factor IX (nine). It is infused into a vein over a period of time.

DDAVP or desmopressin, p.21:

A type of medicine that raises a person's own factor VIII (eight) level in the blood, but is not a blood product. It can be used to treat mild hemophilia A and some types of von Willebrand disease.

diluent, p.35:

The liquid that is mixed with factor concentrate powder. The final product is injected to treat bleeds.

elevation, p.20:

In first aid, elevation means to raise the injured part of the body above the level of the heart. This helps stop bleeding and swelling.

factor concentrate, p.21:

A type of treatment that replaces the missing factor VIII (eight) or IX (nine) by injection into a vein. Factor can be made from human blood plasma and then dried to a powder. This is dissolved in diluent before injection. See recombinant for description of another kind of factor concentrate.

forearm, p.15:

The area of the arm between the wrist and the elbow, a common area for muscle bleeds.

gene, p.5:

Genes carry messages about the way the cells of the body work. For example, they determine a person's hair and eye colour. Hemophilia is passed on through a person's genes.

hemophilia, p.3:

A term used to describe bleeding disorders in which clotting factor VIII (eight) or IX (nine) in a person's blood plasma is missing or is at a low level.

hemophilia A, p.4:

A bleeding disorder in which clotting factor VIII (eight) in a person's blood plasma is missing or is at a low level.

hemophilia B, p.4:

A bleeding disorder in which clotting factor IX (nine) in a person's blood plasma is missing or is at a low level.

hemophilic arthritis, p.12:

Inflammation of a joint, usually with pain and swelling, due to repeated bleeds. Cartilage in the joint breaks down and some bone wears away. Sometimes the joint cannot move.

Glossary

home therapy, p.33:

Injection or infusion with a treatment product away from the hospital, usually at home.

infusion, p.33:

Injection of a treatment product into a vein.

inhibitor, p.23:

Antibodies made by the body to fight off things it sees as foreign which inhibit a blood clotting factor.

IX (nine), p.3:

A Roman numeral that means "9" (nine). Each clotting factor in blood plasma is named with a Roman numeral.

joint, p.9:

The place where two bones meet.

joint capsule, p.9:

The area or sleeve that holds the bones together in a joint (where two bones meet).

mild hemophilia, p.4:

A disorder caused by factor VIII (eight) or IX (nine) activity of 5% to 40% of the normal level in the blood.

moderate hemophilia, p.4:

A disorder caused by factor VIII (eight) or IX (nine) activity of 1% to 5% of the normal level in the blood.

muscle spasm, p.14:

Painful tightening of a muscle that a person cannot control.

nerve, p.14: The sensitive, cordlike fibres that pass messages through the body, including pain.

NSAIDS or non-steroidal antiinflammatory drug, p.29:

A drug, such as ibuprofen, that reduces pain and fever but does not contain steroids.

plasma, p.2:

Part of blood that contains fibrin and clotting factors.

platelets, p.2:

The blood cells that make a plug to patch holes in arteries, veins, and capillaries.

psoas muscle, p.15:

A muscle near the groin that help move the hip joint and the spine.

recombinant, p.21:

A type of factor concentrate that is manufactured in a laboratory instead of being separated from human blood. Recombinant proteins are copies of certain kinds of proteins found in human blood plasma.

severe hemophilia, p.4:

A disorder caused by factor VIII (eight) or IX (nine) activity in the blood, usually less than 1% of the normal level.

spontaneous bleeding, p.8:

Bleeding that happens for no clear reason (not after an injury or surgery).

synovium, p.9:

The lining of the joint capsule. It is made of special cells that make a slippery, oily fluid that helps the joint move easily.

thigh, p.15:

The area of the leg between the hip and the knee which is a common area for muscle bleeds.

upper arm, p.15:

The area of the arm between the shoulder and elbow, a common area for muscle bleeds.

vein, p.1:

A tube or blood vessel that carries blood through the body to the heart. The body has many veins.

venepuncture, p.34:

Putting a needle into a vein. This can be done to take blood or to give an intravenous injection.

VIII (eight), p.3:

A Roman numeral that means "8" (eight). Each clotting factor in blood plasma is named with a Roman numeral.