

What is hemophilia?



WORLD FEDERATION OF
HEMOPHILIA
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What is hemophilia?

- Hemophilia is a bleeding problem. People with hemophilia do not bleed any faster than normal, but they can bleed for a longer time. Their blood does not have enough *clotting factor*. Clotting factor is a protein in blood that controls bleeding.
- Hemophilia is quite rare. About 1 in 10,000 people are born with it.
- The most common type of hemophilia is called *hemophilia A*. This means the person does not have enough clotting factor VIII (8). A less common type is called *hemophilia B*. This person does not have enough clotting factor IX (9). The result is the same for hemophilia A and B; that is, they bleed for a longer time than normal.

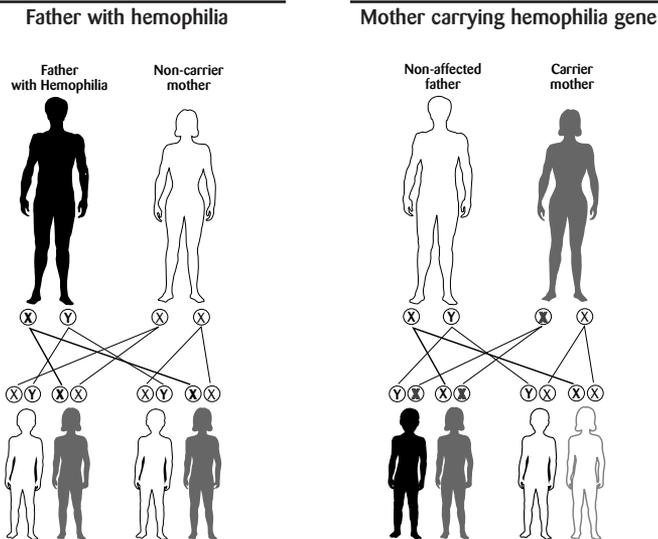
How do people get hemophilia?

- People are born with hemophilia. They **cannot** catch it from someone, like a cold.
- Hemophilia is usually inherited, meaning that it is passed on through a parent's *genes*. Genes carry messages about the way the body's cells will develop as a baby grows into an adult. They determine a person's hair and eye colour, for example.
- Sometimes hemophilia can occur when there is no family history of it. This is called *sporadic* hemophilia. About 30% of people with hemophilia did not get it through their parent's genes. It was caused by a change in the person's own genes.



How is hemophilia inherited?

- When the father has hemophilia but the mother does not, none of the sons will have hemophilia. All of the daughters will carry the hemophilia gene.
- Women who have the hemophilia gene are called *carriers*. They sometimes show signs of hemophilia, and they can pass it on to their children. For each child, there is a 50% chance that a son will have hemophilia and a 50% chance that a daughter will carry the gene.
- Women can only have hemophilia if their father has hemophilia and their mother is a carrier. This is very uncommon.



There are three levels of severity.

The *severity* describes how serious the problem is.

The level of severity depends on the amount
of clotting factor that is missing
from a person's blood.

NORMAL = 50% - 150%
of the normal activity of clotting
factor VIII (8) or IX (9) in the blood.

**MILD
HEMOPHILIA**

5% - 30%
of normal clotting
factor activity

- Might bleed for a long time after surgery or a very bad injury.
- Might never have a bleeding problem.
- Do not bleed often.
- Do not bleed unless injured.

**MODERATE
HEMOPHILIA**

1% - 5%
of normal
clotting factor
activity

- Might bleed for a long time after surgery, a bad injury or dental work.
- Might bleed about once a month.
- Rarely bleed for no clear reason.

**SEVERE
HEMOPHILIA**

less than 1%
of normal
clotting factor
activity

- Bleed often into the muscles or joints (mainly knees, elbows and ankles).
- Might bleed one or two times per week.
- Might bleed for no clear reason.

What are the signs of hemophilia?

The signs of hemophilia A and B are the same.

- Big bruises;
- Bleeding into muscles and joints, especially the knees, elbows, and ankles;
- *Spontaneous* bleeding (sudden bleeding inside the body for no clear reason);
- Bleeding for a long time after getting a cut, removing a tooth, or having surgery; and
- Bleeding for a long time after an accident, especially after an injury to the head.

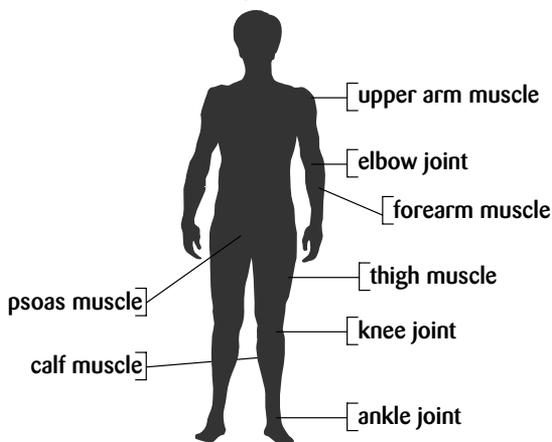
Bleeding into a joint or muscle causes:

- An ache or “funny feeling”;
- Swelling;
- Pain and stiffness; and
- Difficulty using a joint or muscle.

Where does bleeding occur most often?

- People with hemophilia can bleed inside or outside the body.
- If bleeding occurs many times into the same joint, the joint can become damaged and painful.
- Repeated bleeding can cause other health problems like arthritis. This can make it difficult to walk or do simple activities. However, the joints of the hands are not usually affected in hemophilia (unlike some kinds of arthritis).

**Bleeding occurs most often
inside these joints or muscles.**



How is hemophilia treated?

- Treatment for hemophilia today is very effective. The missing clotting factor is injected into the bloodstream using a needle. Bleeding stops when enough clotting factor reaches the spot that is bleeding.
- **Treat bleeding quickly!**
Quick treatment will help reduce pain and damage to the joints, muscles, and organs. If bleeding is treated quickly, less blood product is needed to stop the bleeding.
- **If in doubt, treat!**
If you think you have a bleed, get treatment even if you are not sure. NEVER wait until a joint is hot, swollen, and painful. Do not worry that you may “waste” a few treatments.
- There is no cure yet, but with treatment people with hemophilia can live normal healthy lives. Without treatment, people with severe hemophilia may find it difficult to go to school or work regularly. They might become physically disabled and have trouble walking or doing simple activities – or die young.

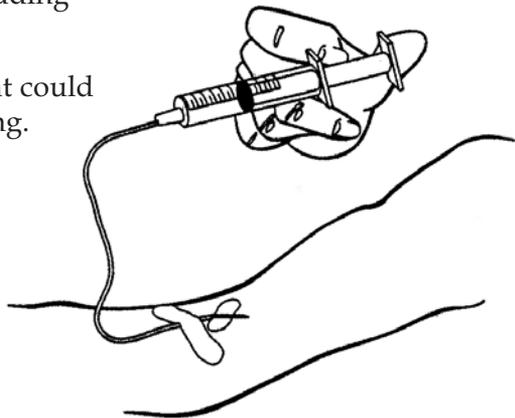
When should treatment be given?

Treatment is given for:

- Bleeding into a joint;
- Bleeding into a muscle, especially in the arm or leg;
- Injury to the neck, mouth, tongue, face or eye;
- Severe blows to the head and unusual headache;
- Heavy or persistent bleeding from any site;
- Severe pain or swelling in any site;
- All open wounds requiring stitches; and
- Following any accident that may result in a bleed.

Treatment is given before:

- Surgery, including dental work
- Activities that could cause bleeding.



When is treatment probably not needed?

- **Small bruises** are common in children with hemophilia, but they are **not** usually dangerous. However, bruises on the head might become serious, and should be checked by a hemophilia nurse or doctor.



- **Small cuts and scratches** will bleed for the same amount of time as in a normal person. They are **not** usually dangerous.
- **Deeper cuts** will often — but not always — bleed longer than normal. The bleeding can usually be stopped by putting direct pressure on the cut.
- **Nosebleeds** can usually be stopped by putting pressure on the nose for five minutes. If bleeding is heavy or does not stop, treatment is needed.

Important Points for People Living with Hemophilia

1. Treat bleeding quickly!

When you stop bleeding quickly, you have less pain, and less damage to joints, muscles, and organs. Also, you need less treatment to control the bleeding.

2. Stay fit.

Strong muscles help protect you from joint problems and *spontaneous* bleeding (bleeding for no clear reason). Ask your hemophilia doctor or physiotherapist which sports and exercises are best for you.

3. Do not take ASA (Aspirin®).

ASA (acetylsalicylic acid) can cause more bleeding. Other drugs can affect clotting, too. Always ask your doctor which medicine is safe.

4. See a hemophilia doctor or nurse regularly.

The staff at a hemophilia clinic or treatment centre will give help and advice about managing your health.

5. Avoid injections into your muscles.

A muscle injection could cause painful bleeding. However, vaccinations are important and safe for a person with hemophilia. Most other medications should be given by mouth or injected into a vein rather than into a muscle.

6. Take care of your teeth.

To prevent problems, follow the advice of your dentist. Dental injections and surgery can cause major bleeding.

7. Carry medical identification with information about your health.

A special international medical card is available from the World Federation of Hemophilia. Some countries sell tags called “Medic-Alert” or “Talisman” that you wear around your neck or wrist.

8. Learn basic first aid. Quick aid helps manage bleeding.

Remember that very small cuts, scratches, and bruises are usually **not** dangerous. They do not usually need treatment. First aid is often enough.



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