

4 Management of Bleeds

This chapter provides information on how to manage bleeds. It includes the following sections:

- An introduction to bleeds
- Bruises
- Mouth bleeds
- Muscle bleeds
- Joint bleeds
- Life-threatening bleeds
- Other types of bleeds

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MYTH

PEOPLE WITH HEMOPHILIA BLEED PROFUSELY FROM MINOR CUTS.

REALITY

Bleeding from minor cuts in people with hemophilia is easily controlled. A little pressure and an adhesive bandage usually does the trick.

Clotting of blood involves a number of stages. The blood of people with hemophilia functions normally in the first three stages—the constriction of the blood vessels, the adhesion of platelets at the site of the injury and the aggregation of other platelets and proteins to plug the hole. These three steps are usually sufficient to stop bleeding from minor cuts. It's only the fourth stage—the formation of fibrin from the chain reaction of clotting proteins like factor VIII and IX—where people with hemophilia run into problems. This coagulation cascade is especially important to stop internal bleeding.

AN INTRODUCTION TO BLEEDS

This chapter describes common bleeds that can happen in children with hemophilia. It will help you learn how to recognize early signs of a bleed so that you can treat it right away. It explains how to treat bleeds, and how to identify serious bleeds that need to be treated at a hospital.

The chapter also describes the types of bleeds that are life-threatening — these bleeds happen very rarely but parents need to learn how to recognize the signs so that they can take their child to the hospital right away. There are also guidelines on First Aid for bleeds, which is an important part of treatment along with clotting factor therapy.

■ What is a bleed?

The word “bleed” means to lose blood from the body due to injury or illness. In the hemophilia context, a bleed refers to abnormal or prolonged bleeding. The medical term for bleed is *hemorrhage*.

Many people with hemophilia experience bleeds because their blood does not produce enough of a certain clotting factor protein — *factor VIII* (hemophilia A) or *factor IX* (hemophilia B). How often bleeds occur will depend on the severity of the person's hemophilia, his treatment plan, and his overall physical health and lifestyle.


Most bleeds in the first two years of childhood are surface bruises resulting from falls and bumps as the baby learns to crawl and walk. Bleeding into *joints* and *muscles* are common features of hemophilia and can occur starting at a very young age, in the preschool years. Internal bleeds are a serious concern — any injury involving the head, neck, chest or abdomen can be life-threatening and therefore requires immediate medical attention.

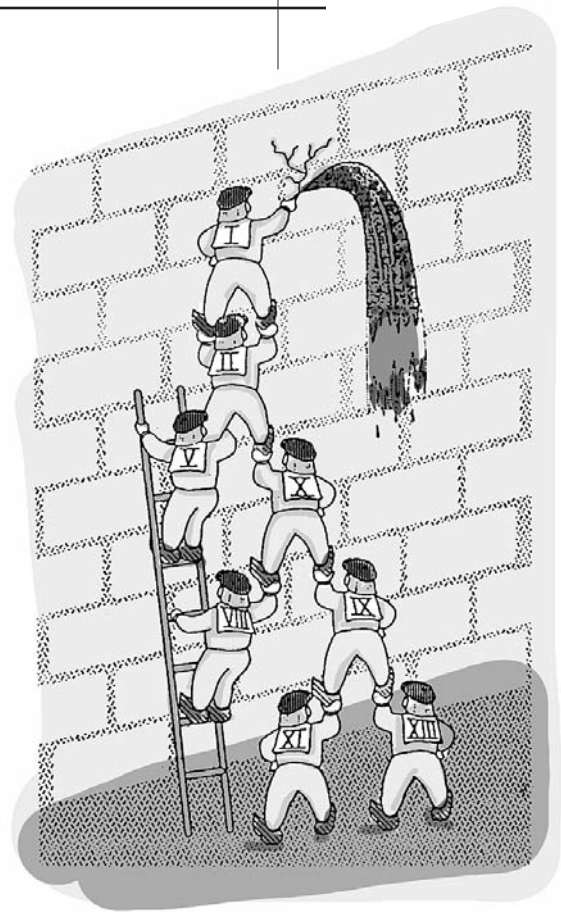
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■ How does a bleed happen?

A bleed happens when blood vessels are torn. Blood then leaks out into the surrounding tissues. In a person with normal clotting, the bleeding stops quickly because *platelets*, tiny cells less than 1/10,000 of a centimetre in diameter, stick to the blood vessel walls. The clotting factors then work to form a substance called *fibrin* to seal the tear in the blood vessel wall. A person who lacks factor VIII or IX will keep bleeding because the clotting factor is not present in sufficient quantity to make fibrin. The blood will clot eventually, but it takes longer and the clot is not as strong.

 For more information on how blood clots, see **Chapter 1, An Introduction to Hemophilia.**



■ How are bleeds treated?

All joint and muscle bleeds, and serious bleeding into soft tissues, must be treated with *clotting factor concentrates*. For some people with mild or moderate hemophilia A, treatment with *desmopressin acetate* (DDAVP) can be enough to stop bleeding. First Aid with rest, ice, compression and elevation — known as **RICE** — helps slow bleeding. **RICE** is also important while a bleed heals, which can take several days or even weeks. Physiotherapy is often part of treatment for bleeds.

“My blood can’t plug the holes.”


Still a Mystery...

The mad monk Rasputin was able to relieve some of the symptoms of hemophilia for the Tsarevich Alexei, heir to the throne of Russia in the early 1900s. It is said he used hypnosis. Did the hypnosis slow or stop the bleeding? Did it relieve the pain? Did it merely calm the boy so that the treatment seemed to help? Or was it a combination of all three? It's still a mystery.

Clotting Factor Concentrates

Factor concentrate is given to replace the deficient factor (factor VIII in hemophilia A and factor IX in hemophilia B) and raise it to within normal levels. This helps the blood clot normally.

- **On-demand therapy** – Clotting factor concentrate is infused as soon as possible after a bleed starts in order to stop the bleeding. In general, joint and muscle bleeds are treated this way. *Clotting factor therapy* (also called factor replacement therapy) is continued until the bleeding has stopped and the risk of re-bleeding is reduced. In some cases, especially when treatment is given soon after bleeding starts, one infusion is enough. More frequent infusions of factor concentrate (two to three times per day) are given to treat more serious bleeds. A serious injury can require frequent infusions for several days or even weeks to prevent re-bleeding and make sure the bleed heals completely.
- **Prophylaxis therapy** – Preventative treatment in hemophilia is called *prophylaxis*. Factor concentrate is infused one or more times a week to help prevent bleeds. Many children with moderate or severe hemophilia in Canada are treated with prophylaxis to help prevent bleeding from occurring at all. Prophylaxis therapy is one part of prevention. Maintaining healthy muscles and joints through regular physical activity and by selecting activities that do not cause bleeds is also important.

 For more information, see: **Chapter 6, The Role of Prophylaxis; Chapter 8, Complications of Hemophilia – Joint and Muscle Damage; and Chapter 12, Physical Activity, Exercise and Sports.**

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Desmopressin Acetate (DDAVP)

Desmopressin acetate (DDAVP) is a synthetic hormone that is helpful in the treatment of the majority of people with mild hemophilia A, but not all. Therefore, individuals should be tested to see how well they respond to DDAVP. The use of desmopressin will depend on the severity of the bleed. If the bleed is serious or life-threatening, treatment with clotting factor concentrates must be given. DDAVP works by raising the factor VIII levels. It only works for hemophilia A — it is not effective at all for hemophilia B.

 For more information on desmopressin, see **Chapter 5, Clotting Factor Therapy**, and **Chapter 9, Mild and Moderate Hemophilia**.

First Aid (Rest, Ice, Compression and Elevation)

Bleeding can be *slowed* by using First Aid steps called **RICE** — this stands for **Rest, Ice, Compression** and **Elevation**. These steps can be used for a small bleed into soft tissue or a *superficial muscle* (an outer muscle as opposed to a deep or inner muscle). **RICE** is also used for joint and muscle bleeds to help reduce pain and swelling while the clotting factor concentrate is being prepared for infusion and during the recovery period.

The HTC team will teach you when to apply RICE and how to apply each step. If you are at all unsure about whether to apply RICE or for how long, be sure to consult the physiotherapist for advice.

REST

When a bleed occurs and while it is healing, the affected limb should be rested. This means that if your child has an elbow or shoulder bleed, he should rest his arm, not move it around and not use it to lift or carry things. If he has a joint or muscle bleed in his leg, walking should not be allowed — he should be kept off his feet as much as possible. It should be rested until it heals — until it has fully returned to its state of health and physical condition before the bleed.

R
Rest

I
Ice & Immobilization

C
Compression

E
Elevation



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ICE

Apply ice to an injured area to reduce pain and muscle spasms. Ice also helps reduce swelling and redness, also referred to as *inflammation*. You can use an ice pack, crushed ice in a plastic bag or a bag of frozen vegetables wrapped in a damp towel. There are many different ways to apply ice, but it is very important not to apply the ice for too long.

Important points to remember about using ice include:

- Do not use ice over open cuts or scrapes.
- Do not use ice if there is poor circulation or poor sensation in the injured area.
- Be sure your child can tolerate the ice. Some young children may feel that the ice is more uncomfortable than the bleed.
- Do not leave the ice on longer than the recommended time. Leaving ice on too long (more than 15 minutes) can cause muscle weakness and can also cause an increase in blood flow.

There are many ways to apply ice to an injury to help decrease pain and swelling:

- **Ice Packs** – Use crushed ice (or a bag of frozen peas or corn) wrapped in a damp towel. Wrap the towel and ice firmly around the injured area. (Large ice cubes are not as easy to ‘fit’ around a joint.) Apply the ice for 10 to 15 minutes.
- **Cold Packs (gel packs, chemical cold packs)** – Wrap the pack in a thin damp towel. Make sure the pack is flexible and mold it to fit the area to be treated. Never put the cold pack directly on the skin. Leave the cold pack on for 10 to 15 minutes.
- **Ice Cups/Ice Massage** – Fill small paper cups or popsicle makers with water and keep them in your freezer until they are needed. Rub the ice directly over the injury, in smooth strokes up and down or in circles. Have an extra towel close by to catch the drips. Massage lightly for 3 to 5 minutes, or until the skin feels numb. This way of using ice is very cold.
- **Ice Bath** – Add ice cubes to a bucket of cold water. Put the injured limb in the water for 1 to 5 minutes as tolerated. This works well for a bleed in the ankle, foot, wrist or hand but it is also very cold and may not be tolerated well by young children.



“It’s not easy to hold an ice pack against the forehead of a screaming 2 year old.”

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IMMOBILIZATION

The **I** in **RICE** can also mean **Immobilization**. This involves using a temporary half-cast or splint to limit movement. It helps rest the injury and prevents the child using an injured arm or putting weight on an injured leg.

Important points to remember about immobilization:

- A full cast is not recommended. If the swelling continues to increase, the cast will be too tight and blood circulation can be cut off (not enough blood will circulate to the rest of the limb).
- Immobilization should not be used for a long period of time because joints become stiff and muscles become weak if they are not used. The injured joint or muscle must be examined by the HTC physiotherapist every 3 to 5 days to see when the cast or splint can be removed.
- The physiotherapist will also recommend exercises to help restore joint mobility, muscle strength and balance.

Immobilization of a joint with a half-cast or a removable splint is useful when:

- A bleed is severe enough to limit motion.
- There is a lot of pain with movement and at rest.
- A child is too young to understand that he must rest the injured part.

“At least I get to play video games when I have a bleed.”

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COMPRESSION

Put pressure on the injured area to “pinch off” the blood vessels and slow down bleeding. This helps limit swelling to the injured area and keeps it from progressing to other parts of the limb. A joint or muscle bleed is compressed by using an elastic (tensor) bandage or an elastic compressive sleeve such as Tubigrip®, Elastogrip® or Surgigrip®. If the injured joint or muscle is very sensitive, compression might not be tolerated in the early stages.

General guidelines for compression to slow bleeding include:

- Wrap the injured joint or muscle starting well below and ending well above the bleeding area, using a criss-cross pattern.
- Be careful not to wrap the limb too tight in order to avoid a “tourniquet” effect causing blood circulation to be cut off (not enough blood will circulate to the rest of the limb).
- Watch carefully for coolness, numbness or a change in colour to the skin below the wrap and farther parts of the body such as fingers or toes. If any of these symptoms occur, remove the bandage and re-wrap it less tightly.
- An alternative is a compressive sleeve (Tubigrip™, Elastogrip™ or Surgigrip™), which many parents find easier to use, especially with young children.
- Use extra padding directly over the bleed (this is particularly useful for thigh bleeds).

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ELEVATION

Raise the injured arm or leg to a level higher than the heart (for example, by using cushions or pillows) to help decrease blood pressure and slow the bleeding. Elevation also helps reduce “tracking” of swelling along an injured limb.

A joint or muscle bleed is never minor. Bleeding into a joint or muscle will cause pain, swelling and loss of movement. If the bleeding continues, it can cause serious damage. First Aid (**RICE** – rest, ice, compression and elevation) is important – but factor concentrate treatment is absolutely essential. Factor should be infused as soon as possible to stop the bleeding. Sometimes, it can be hard to judge how serious a bleed is and whether factor concentrate is needed. Follow the HTC motto:

“When in doubt, treat and infuse.”

Physiotherapy and Exercises

Initial treatment of a joint or muscle bleed using **RICE** and factor replacement therapy should generally be followed by physiotherapy and exercises. This is important to restoring joint and muscle strength, function and range of motion. The HTC physiotherapist will work with you to set up a suitable exercise program for your child. The physiotherapist will also supervise his progress and adjust his physiotherapy program accordingly.

 For more information on physiotherapy and exercises, see **Chapter 12, Physical Activity, Exercise and Sports**.

“Be careful not to use pain level as the only guideline for deciding when he can return to his regular physical activities. Make sure he has fully returned to his state of health and physical condition before the bleed.”

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■ Do all bleeds need to be treated with factor?

For a very minor bruise or bleed, the first aid **RICE** steps (rest, ice, compression and elevation) may be enough treatment. The bleed may not necessarily require treatment with clotting factor.

Any bleeding into a joint and significant bleeding into soft tissues, especially a muscle, *must* be treated with clotting factor.

A significant soft tissue or muscle bleed is one that causes pain and limits the movement and function of nearby joints. A deep muscle bleed may not be obvious at first because you don't usually see bruising or swelling right away — but if the bleeding continues, it can cause damage to nerves and blood vessels. It is therefore important to also watch for signs such as pain and loss of movement. Joint and muscle bleeds must be treated with clotting factor as soon as possible. Your care team will help explain how to recognize these bleeds.

Any injury to the head or vital organs can be very serious and needs treatment as soon as possible. An injury or bleed that involves the neck, throat or chest can interfere with the airway for breathing — this can happen very suddenly. It is critical to act fast before this happens. Infuse factor, if possible, and take your child to the hospital without any delay.

■ Can all bleeds be treated at home?

Once you have learned how to infuse clotting factor, you will be able to treat most joint, muscle, mouth and nose bleeds at home. However, it is important to remember that even though you will be taking charge of your child's home infusion program, the HTC members are still a vital part of his care team.

You must keep the care team informed about every serious bleed so that they can help you decide whether you need to take him to the HTC or nearby hospital to be seen by a doctor. If a visit to the

“You must keep accurate home infusion records and communicate regularly with the HTC about your child's bleeds and treatment. Never hesitate to discuss any concerns or problems with the care team.”

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hospital is not needed, the care team can give you advice on using **RICE** and/or clotting factor to stop the bleed. The physiotherapist can guide you on how to help your child gradually restore strength and function to the joint and/or muscle.

Injuries to the head, neck, chest or abdomen are very serious. For these types of bleeds, you need to give clotting factor as soon as possible *and* take your child to the nearest hospital right away. Contact your HTC after you get to the hospital.

BRUISES

■ How should bruises be treated?

A bruise is caused by bleeding into the tissues that does not break through the skin — this results in pain, swelling and redness. The medical term for a bruise is *hematoma*. Treatment depends on a number of factors such as the site and size of the bruise, and how much discomfort or pain it causes. Although bruises can look alarming, they may not always require treatment with factor. Ice and compression is sometimes enough to reduce swelling or pain from a minor bruise.

Your hemophilia treatment team may suggest treatment with clotting factor if the bruise...

- is very painful (for example, a buttock bleed)
- is getting larger in size
- limits movement in nearby joints
- is located near a critical place (for example: eyes, throat, major artery or nerve, etc.)

Sometimes as a bruise starts to heal, a small bump or “knot” can be felt just under the skin. This is normal and will go away as the mass of blood under the skin is re-absorbed. Large bruises may take several weeks to heal completely.

MOUTH BLEEDS

■ What are common causes of bleeding in the mouth?

A child with hemophilia can bleed from the mouth in several ways...

- He can accidentally bite his tongue.
- He can bite the gums on the inside of his cheeks.
- He can bleed when baby teeth fall out or after tooth extractions.
- He can get a cut in his mouth from eating food with sharp edges such as potato chips or from poking a sharp object such as a pen or pencil in his mouth.

■ Do cuts in the mouth bleed a lot?

Yes, they can. Bleeding in the mouth can be troublesome and messy and the blood mixed with saliva sometimes makes it look worse than it is. Serious cuts in the mouth, however, may bleed for a long time.

In some cases, bleeding may stop and then restart after a few hours or even days. There are several reasons for this. First, the mouth's moistness makes it harder for a clot to form and stay in place. Second, saliva can dissolve or wash away a protective blood clot. Third, movement of the tongue or vigorous chewing can dislodge the clot.

“Tongue bleeds can be very difficult. I didn't expect the cut to keep reopening.”

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The amount of blood lost from a cut in the mouth can sometimes be underestimated if a lot of the blood is swallowed. Signs of this include:

- pale skin
- lethargy (tiredness, lack of energy)
- nausea
- upset stomach
- loss of appetite
- vomiting blood
- dark stools

Therefore, it is important that you have your child checked at the HTC whenever there is a persistent mouth bleed.

■ How should a mouth bleed be treated?

The following treatment plan is recommended for a mouth bleed:

- Apply firm pressure to the site of the bleed. Use a popsicle or ice to help slow bleeding and relieve pain.
- Infuse clotting factor if 20 minutes of pressure and ice have not stopped the bleeding.
- Give your child only soft foods (yogurt, pudding, custard, etc.) and avoid giving crunchy and chewy food until the injury heals.
- Avoid giving hot fluids and hot food for several days to a week, until the injury heals.
- Babies should drink from a cup rather than a bottle and not use a pacifier, if possible. Children should not use drinking straws until the injury heals.


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Your doctor may prescribe tranexamic acid (Cyklokapron®), which can be taken orally in pill or liquid form. Tranexamic acid should not be given if your child has *hematuria* (blood in the urine).

If the bleeding seems serious or does not stop after treatment, your child should be seen at the HTC or by his pediatrician. Serious cuts to the lip or gums may need to be treated with clotting factor — if not, a mouth bleed can sometimes ooze for several days. Persistent bleeding can cause severe *anemia* — the person's red blood cell count will be lower than normal.

■ What can be done to prevent mouth bleeds?

Never let your child run with anything hard in his mouth, and always insist that he sit down to eat. Good dental care is very important to keep gums healthy and prevent bleeding and gum disease. Regular brushing and flossing of teeth should be started at an early age.

 *For more ideas on prevention, see Chapter 11, Staying Healthy.*

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MUSCLE BLEEDS

■ What causes a muscle to bleed?

A muscle is a bundle of tissue and fibers that have the ability to contract. This produces movements such as lifting, standing up and sitting down, and walking. A bleed in a muscle can occur when it is over-exerted or over-stretched, or when there is a direct blow to it. This causes muscle fibres to tear and bleed.

■ How will I know if my child has a bleed in a muscle?

Some muscle bleeds can be tricky to detect. The blood can seep between the layers of muscle and not cause swelling right away. Sometimes, this is mistaken as a strained muscle. But as the bleeding continues, it will become painful and the muscle may tighten or weaken. Movement of the limb or joint near the muscle bleed will become limited because it will hurt to flex or stretch the muscle.

Signs and symptoms of a muscle bleed include:

- Your child holds a part of his body in an awkward position or seems reluctant to use it.
- He complains of pain or a tingly sensation in the injured area.
- The injured area feels warm, swollen and/or firm to the touch.

It is not common to see bruising with a muscle bleed — if you see any of these signs, infuse clotting factor right away.

“One morning I went to pick my son up from his crib. He was awake and looked OK. As soon as I picked him up, he started to cry as if he had a lot of pain. I pulled off his pajamas and couldn’t see any problem. At the clinic, the care team showed me how to isolate the area that was bothering him. It was his thigh muscle that was the problem. I couldn’t determine this at first. I learned a lot that day and actually felt more confident.”

“Around age 6 or 7, children often have enough coordination and strength to use crutches. Depending on the severity of your child’s hemophilia and other factors, the HTC physiotherapist may recommend that a set of crutches be kept at home in case he has a joint or muscle bleed in his leg. She/he will teach him how to use them properly.”



■ What are the steps for treating a muscle bleed?

Bleeding into a muscle needs to be treated if there is pain that limits movement. Once you have recognized bleeding into a muscle, these steps should be taken as soon as possible:

Replacement therapy

Clotting factor may need to be given daily for a few days or longer, depending on the bleed. Follow the HTC instructions.

RICE

- **Rest** – Keep your child off his feet (in the case of a leg bleed) or restrict his arm movement (in the case of an arm bleed). Complete bed rest may be needed depending on the seriousness of the bleed.
- **Ice** – Apply ice to the muscle to temporarily help reduce pain and muscle spasm.
- **Immobilization** – If the muscle bleed is very painful, the arm or leg may need to be temporarily supported in a splint or half cast. This is done after bleeding stops. A splint will be adjusted over time to gently stretch the muscle back to its normal length.
- **Compression** – Wrap the injured muscle firmly using a tensor bandage or a compressive sleeve (Tubigrip™, Elastogrip™ or Surgigrip™) to help control swelling and pain and help the muscle heal. In some situations, foam padding is used to provide more pressure.
- **Elevation** – Keep the arm or leg elevated on pillows to help reduce swelling.

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Physiotherapy

As a muscle bleed heals, scar tissue can form. The muscle can also become weak after a large bleed. There is risk of an early re-injury or re-bleed if the person returns to his normal activities too soon. The physiotherapist will show your child exercises to progressively stretch and strengthen the muscle, and tell you when he can return to his regular activities.

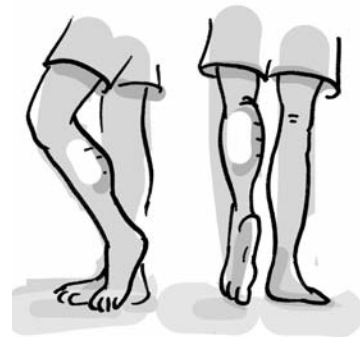
■ Are muscle bleeds serious?

Yes they can be. If an injured muscle is not stretched back to its former length, it can affect the nearby joints. The muscle *and* the joint may have a higher risk of bleeding.

A muscle bleed can also be serious because it can cause damage to important nerves and blood vessels. This happens when there is a *compartment bleed*, a deep muscle bleed inside a closed-in space. A large bleed in these compartments can put pressure on nerves and blood vessels, and cut off blood supply to the muscle. If this pressure lasts long, the damage can be permanent.

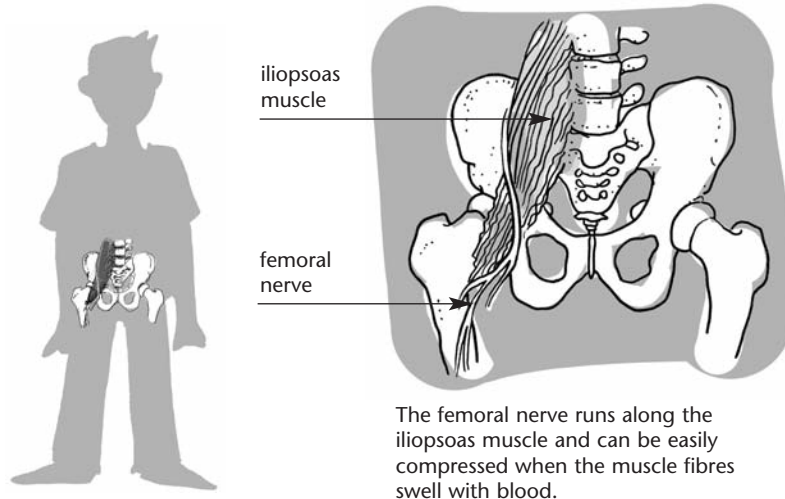
Compartment bleeds can occur in several muscles:

- **forearm muscles** – Bleeding into the deep muscles of the forearm can put pressure on the nerves and blood vessels going to the hand.
- **calf muscle** – A bleed into the calf muscle behind and below the knee can affect the nerves going to the ankle. Sometimes, the person is not able to take full footsteps with the injured leg and walks on his toes instead to avoid painful stretching of the muscle.



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- **iliopsoas** (pronounced ee-lee-o-so-as) **muscle** – This is a large muscle in the pelvic region near the hip joint. Classic signs of this type of muscle bleed are the inability to straighten the hip fully or to stand with the leg and back straight. Bleeding into this muscle is serious and can damage the large nerve that controls the muscles at the front of the thigh.



Factor replacement therapy needs to be given immediately. If your child feels numbness along the thigh and/or has difficulty straightening his knee, it could be a sign of nerve pressure — call the HTC right away for instructions on the treatment needed.

Children with a bleed in the iliopsoas are sometimes admitted to hospital for treatment. An ultrasound scan can help determine the initial size and location of the bleed, and how well it is healing over time. This muscle can take a very long time to recover from a bleed. Total bed rest or use of a wheelchair is required at first — crutches are not helpful early on because the effort of keeping the injured leg off the ground strains the muscle.

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The physiotherapist will guide you on how to protect the injured iliopsoas muscle in the initial stages and suggest gradual exercises to help the muscle's full recovery.

If your child experiences numbness, pain or a tingling “pins and needles” feeling around an injured muscle or in his fingers or toes, it could be a sign of nerve or blood vessel compression. Call the HTC immediately.

■ How can muscle bleeds be prevented?

Muscles can be protected from direct types of injury that can occur with many sports by using the right kind of padding for different activities. (For example: padded shorts or pants for skiing and skating, chest protection for volleyball and shin pads for soccer.) Sometimes muscle bleeds can occur when a person starts doing a challenging new sport or activity without the right preparation to strengthen and train his body and muscles. Proper stretches and warm-ups before sports are very important.

Despite these measures, it is impossible to prevent all muscle bleeds. Therefore, it is also important to make sure that a muscle bleed is *completely* healed before the person returns to his regular activities. A muscle is at risk of getting injured again and re-bleeding if it has not regained its full strength and flexibility. The physiotherapist will guide you on how and when your child can return to his usual activities.

“It is important to remember that most bleeds need to be treated with RICE and clotting factor. Be sure to consult the HTC physiotherapist about how to gradually restore strength and function to a joint and muscle, and when it is safe for your child to return to his normal activities.”

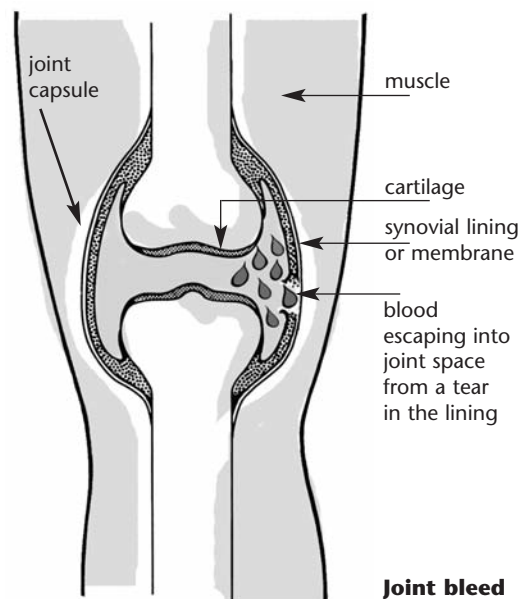
JOINT BLEEDS

■ What causes a joint to bleed?

A joint is the place where two bones come together, allowing movement such as bending, rotating and swinging back and forth. The end of each bone is protected by *cartilage*, and the bones and muscles are held together by connective tissues called tendons and ligaments.

The joint space is surrounded by a thin layer of tissue (membrane) that forms a “sleeve” around the joint — together this is called the *joint capsule*. The joint capsule is lined with a layer of special cells, called the *synovium* or *synovial membrane*. These cells produce small amounts of fluid that make it easier for the bones to move. The synovium also contains a network of small blood vessels that brings nutrients and oxygen to the joint.

The synovium can be torn if it is pinched when a joint moves the wrong way. When there is a tear in the synovium, blood escapes from the blood vessels and begins to fill the joint. In a person with normal clotting, the bleeding stops and the person does not realize he has had a bleed. In a person with hemophilia, the bleeding takes a long time to stop by itself because his blood does not produce enough clotting factor. As the bleeding continues, the joint area begins to swell. Bleeding into a joint is called a *hemarthrosis*.



“Our first joint bleed was traumatic. It felt like it was happening to us. It was very painful. I made sure to have Tylenol with codeine in the house after that. We were all exhausted from the stress of having to watch our son endure such pain. It changed our idea of what hemophilia was.”

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■ Why do some joints bleed more than others?

Joints such as the **knees**, **ankles** and **elbows** (unlike the hip and shoulder) can only bend and straighten in two directions, and cannot easily twist or bend side to side. These joints are also not as well protected by muscle as some of the other joints. For these reasons, they are more vulnerable to physical stress and injury, and bleed most often. Knee, ankle and elbow bleeds tend to develop as children start to do more physical activities. Other joints such as the wrists, fingers and toes can also have bleeds but are less common.

Joints like the **hips** and the **shoulders** are formed like a ball and socket. They can move in many directions, and are protected by strong muscles. While bleeds do happen in these joints, they are not frequent. However, it is important to remember that a bleed into the hip joint can be serious. Bleeding into the hip joint can cause pressure on the main artery that supplies blood to the head of the femur (the 'ball' part of the thigh bone that fits into the hip 'socket').

If a bleed is suspected in the hip area (symptoms include pain, limited motion of the hip and difficulty putting weight on the affected leg), contact your HTC and have your child assessed as soon as possible.

■ How will you know if your child has a joint bleed?

The HTC physiotherapist will help you understand how different joints move so that you will be able to tell when there is a problem.

When your child is still a baby and not yet able to tell you what is wrong, you have to watch out for telltale signs.

“ We check our son over carefully, from head to toe, every evening at bath time. He doesn't even know we are checking him. But we look for bruises and swelling, and make sure he can move his joints all the way. Our physiotherapist showed us how to do this. It's a lot easier if we find something and treat it right away than to have to deal with it in the middle of the night. ”

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MYTH

PEOPLE WITH SEVERE HEMOPHILIA EXPERIENCE SPONTANEOUS BLEEDS; THAT IS, BLEEDS THAT START FOR NO REASON.

REALITY

Physicians and physiotherapists now believe there is always a reason for a bleed, even if it is not apparent to the individual with hemophilia. The cause could be a very slight impact, stretch or twist. Joints are especially prone to bleeding in the presence of an enlarged synovium.

- He is cranky or fussy because of discomfort or pain.
- He does not move an arm or leg as he usually does.
- He cries suddenly when you are changing his clothes or picking him up, or if you try to move the injured limb.

You might sense or notice that something is wrong, but may not know what it is at first. Check him over carefully. Undress him so you can see if there are any signs of a bleed. It is important to keep in mind that bruising is not a reliable way to determine if there is a muscle or joint bleed. It is also important to check that the joints look, feel and move as normal. You may be able to feel warmth over a joint. These are early signs of a bleed. If the bleed continues, there may be swelling.

Once your child starts moving around, you may notice that he is not moving the way he normally does.

- He might not crawl smoothly.
- He may walk or run with a limp.
- He may avoid using one arm, or use his left arm when he normally uses his right.

Check him over carefully, and look for warmth and swelling. Assess his joints by comparing the size, shape and movement on each side of his body.


As your child grows older and has had some experience with joint bleeds, he may describe a “funny” or a “tight” feeling in the joint when a bleed first starts. You may not be able to see that anything is wrong, and you may be tempted to “wait and see.” Remember the HTC team motto:

“When in doubt, treat and infuse.”

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Management of Bleeds

That's because if the bleeding is stopped in the very early stages, it may require fewer infusions and the recovery time will be shorter. Delay in giving clotting factor allows more blood to escape into the joint and means the recovery time will be longer — the risk of permanent damage will also be greater.

 For more information on joint damage, see **Chapter 8, Complications of Hemophilia.**

Sometimes a child will try to hide the fact that he is having a bleed. He may not want to stop playing or may fear missing a special event that he has been looking forward to. He may not like getting his “poke.” Or he might have been injured doing something you have told him not to do! In these cases, you may notice changes in your child's behaviour or movements. For example, he may avoid walking in your presence so that you don't see his limp.

It is up to you to take charge. Take him aside, assess him carefully, and make sure he gets treatment if it is needed. He needs to understand that treatment is not punishment, but a way to get him better and back to his play faster.

■ What are the steps for treating a joint bleed?

Once you recognize bleeding into a joint, these steps should be taken as soon as possible:

Replacement therapy

The most important treatment is immediate infusion with clotting factor. Your HTC will advise you on the amount to infuse. This will depend on ...

- the type and severity of your child's hemophilia
- his weight
- the type of bleed

“Every night when I get ready for bed I put on my pajamas, brush my teeth and check my joints. If I need treatment, I tell my mom.”

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Nicholas:

“You mean—let’s pretend—if a bleed started right now, and I told my mom, and I got my needle right away—you mean, I wouldn’t need a cast?”

Physiotherapist:

“That’s right. The joint wouldn’t have time to swell and get sore, because the bleeding would stop right away.”

Nicholas: *“....and I wouldn’t need to come back tomorrow?”*


Physiotherapist:

“That’s right. The sooner you tell someone and get your poke, the easier it is for everybody.”

Nicholas:

“Wow!”

Certain bleeds require a larger dose of factor concentrate than others. For example, a child with hemophilia A who has a small bleed into a joint needs a dose to bring his factor VIII level to within 20 to 30 percent of normal. However, a severe joint bleed requires a larger dose, one that will bring his factor VIII level to 50 percent of normal.

 For complete information on suggested dosages for different types of bleeds, see Tables 2 and 3 in **Chapter 5, Clotting Factor Therapy**.

RICE


Rest the joint, apply ice, use a tensor bandage or compressive sleeve, and elevate the limb. If the injury is painful and movement is limited, the joint may need to be immobilized for a short period in a splint or half-cast. Your HTC team will advise you if this is necessary.

Physiotherapy

After the bleeding has stopped, physiotherapy and gradual exercises are important. This will help your child *progressively* regain full range of motion of the joint, full muscle strength and very good muscle reaction (reflex), which is important for balance. The HTC physiotherapist will guide you on the steps that need to be followed to help the joint fully recover.

It is important not to increase your child’s physical activity level or let him return to full activities too quickly following a joint bleed.

If there is still pain or joint swelling (post-bleed *synovitis*) — even if swelling is painless or seems insignificant — stress and exertion can cause re-bleeding and lead to a *target joint* and/or joint damage. The physiotherapist will advise you on when your child can return to his usual physical activities.

 For more information on chronic synovitis, see **Chapter 8, Complications of Hemophilia**.

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Management of Bleeds


■ How much clotting factor is needed and for how long?

Inadequate treatment with clotting factor (not enough and/or not enough times) can lead to repeat bleeding into the same joint. For this reason, clotting factor should be used until there is no longer any pain and swelling and full range of motion has returned.

■ How can joint bleeds be prevented?

Absolute prevention of any joint bleed may not be realistic. Nevertheless, there are many steps that can be taken to help reduce how often joint bleeds occur and the severity of bleeds. Many HTCS recommend the use of preventative treatment (prophylaxis), so that the individual will always have levels of clotting factor in his blood that are closer to normal in case of injury.

It is important to regularly consult the HTC physiotherapist about which physical activities and sports are recommended for your child at different ages. Some sports are not recommended because they come with a greater risk of injury and bleeds. Strong muscles, proper footwear, and knee and elbow pads also help to protect joints from injury. Ankle supports can be worn for certain sports or while an ankle heals. Ask the physiotherapist for advice on how to prevent joint bleeds.

 For more information about prevention of bleeds, see **Chapter 6, The Role of Prophylaxis; Chapter 11, Staying Healthy; and Chapter 12, Physical Activity, Exercise and Sports.**

Test Your Knowledge

.....
What do the letters R-I-C-E stand for with regard to hemophilia care?

(For some help in finding the answers, see pages 4 to 8 in this chapter.)

(The correct answers are on page 17-17.)

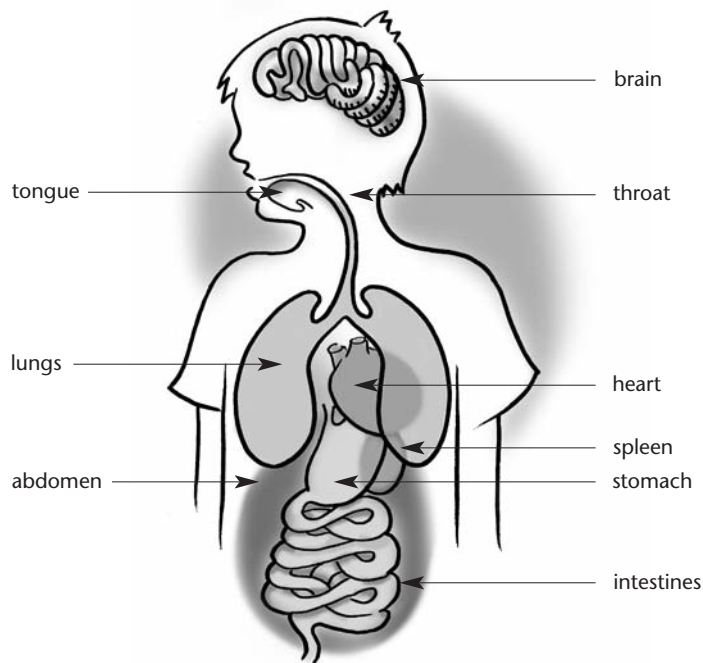
LIFE-THREATENING BLEEDS

Bleeding into the head, neck, chest, or abdomen can be life-threatening. Remember! Sometimes you may not see or recognize the injury when it occurs, especially if the bleeding is internal. Watch for signs such as pain, nausea or difficulty breathing — take your child to the hospital right away if you see any of these symptoms.

HEAD INJURIES

■ What should I know about injuries to the head?

All injuries to the head need to be taken seriously because of the risk of bleeding into the brain. The brain is the control centre for all life-sustaining functions. A bleed into the brain is very serious.



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Management of Bleeds

■ How should head bumps be treated?

Minor head bumps can be frustrating because it's hard to know whether or not to treat with clotting factor. Head bumps are especially common in young children at the toddler stage (ages one to two), as they are just learning to walk and run and are unsteady on their feet. Toddlers often bump into doors, walls and furniture. Many times the child is not upset by the injury — he doesn't even cry — and often there is no bruise or cut.

It may not be necessary to treat simple head bumps with clotting factor if the child does not have any of the symptoms of a serious head injury (described below). If you are not sure, you should speak to the HTC physician or nurse coordinator.

■ How do minor head injuries need to be treated?

Minor head injuries are much more common than major head injuries. A child with a minor head injury may have bruising or a small cut at the site of the head injury, but does not have any of the signs or symptoms listed below. Children with minor head injuries should receive at least one infusion of clotting factor, and must be observed closely for symptoms of a serious head injury. In general, children with minor head injuries do not need to be admitted to hospital, and special X-ray studies are not necessary.



■ How can you tell that a head injury is serious? What do you need to do?

Watch out for any of these signs and symptoms:

- headache
- blurred vision
- nausea or vomiting
- mood or personality changes
- drowsiness
- loss of balance or coordination
- weakness or clumsiness
- stiffness of the neck
- loss of consciousness
- seizures

If any of these symptoms appear, you must take your child to the hospital right away. Serious head injuries require immediate infusion of clotting factor to increase factor levels to 100 percent of normal. If you can, treat your child with clotting factor before leaving for the hospital. Call ahead to your HTC or the hospital. Inform the staff that you suspect a brain hemorrhage and are on route to hospital. This way, they can prepare for your arrival.

People with hemophilia with serious head injuries need to be admitted to hospital. Special X-rays (CT scan or MRI) of the head need to be done to look for bleeding into or around the brain. The person will need to be watched closely. Clotting factor must continue to be given until the doctors are sure that bleeding into the brain has not occurred or that bleeding has stopped completely. If bleeding has occurred, surgery may be necessary to remove the blood that has accumulated.

“Why do most triage nurses not understand how serious a head bleed can be! Parents need to advocate on behalf of their child.”

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Management of Bleeds

■ How can head injuries be prevented?

The way to prevent head injuries is to use good common sense safety precautions. For example, never leave your baby alone on a diaper change table or bed from which he can roll off. Place gates at the top and bottom of all stairs. Some HTC's recommend the use of a helmet as the toddler learns to walk. When he gets older, insist that he wear his helmet when bicycling.



NOSE, MOUTH AND THROAT INJURIES

The tissues in the nose, mouth and throat contain many blood vessels. Injury or infection can result in blood filling these tissues. As these tissues swell with blood, they can press on the airway, making it smaller or closing it completely.

Watch out for:

- pain in the neck or throat
- swelling
- difficulty swallowing
- difficulty breathing

If any of these symptoms appear, you must take your child to the hospital right away.

“My son had a bleed in the throat brought about because of heavy coughing for several days. He swallowed the blood and eventually brought it up. Although it was not a serious bleed, it was very upsetting and frightening until he was x-rayed.”

“As a toddler, my son fell on a railway tie bordering a sand pit at a park. Later in the day I noticed obvious swelling in the abdomen. An x-ray showed he had torn the lining in his stomach, and a bleed had developed. He has moderate hemophilia. He was kept in the hospital overnight for observation. We were distraught at the time. It wasn’t something we had experienced or heard about.”

CHEST INJURIES

The lungs, heart and major blood vessels are located in the chest cavity. Injury to the chest may cause bleeding in vital organs. Bleeding in the lung tissues forces blood into the spaces that normally contain air. This makes breathing difficult.

Watch out for:

- pain in the chest
- difficulty breathing

If any of these symptoms occur, take your child to the hospital right away.

ABDOMEN

The stomach, spleen and intestines are just three of the organs found in the abdomen. Injury to this belly area could result in massive bleeding from an organ or major blood vessel. Without treatment, this could be fatal.

Watch out for:

- pain in the abdomen or lower back
- nausea and/or vomiting

If any of these symptoms occur, take your child to the hospital right away.

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Management of Bleeds

OTHER TYPES OF BLEEDS

NOSE BLEEDS

■ Are nose bleeds common in children with hemophilia?

Not necessarily. Some children with hemophilia will never have a nose bleed. Others, however, will have them once in a while. In a person with hemophilia, nose bleeds, like other bleeds, take longer to stop.

■ How can a nose bleed be stopped?

Children should be taught to calm down as much as possible in the event of a nose bleed. A nose bleed can be stopped by sitting upright and firmly pinching the lower part of the nose (at the cartilage) for 10 to 15 minutes. Some people find that a cold cloth placed on the back of the neck and on the bridge of the nose is helpful.

It may be necessary to keep on pinching the nose for another 10 to 15 minutes. If the nose bleed still does not stop after 20 to 30 minutes, call the HTC to discuss whether treatment with desmopressin or clotting factor is needed. An antifibrinolytic agent (Cyklokapron®) can be given for 5 to 7 days after the nose bleed to prevent re-bleeding.

Drinking hot liquids and strenuous exercise can cause a nose bleed to restart. Therefore, it is helpful to avoid hot soup and hot drinks and vigorous physical activity for 24 hours after a nose bleed.



“It seems the months of November and December are nose bleed months, even though we have bought a good humidifier.”

Test Your Knowledge

.....

Which of the following are signs of a serious head bleed (intracranial bleed)?

- a) headache
- b) blurred vision
- c) a small bruise
- d) nausea or vomiting
- e) a minor cut on the head
- f) mood changes
- g) loss of balance or coordination

(For some help in finding the answers, see pages 25-28 in this chapter.)

(The correct answers are on page 17-17.)



■ What can be done to prevent nose bleeds?

There are several easy ways to prevent or reduce the frequency of nose bleeds. It is important to maintain a certain level of humidity in the house, especially in the child's bedroom. This is especially important in the winter when heating makes a house much drier. A humidifier is ideal but an open bowl of water can also work very well. Spreading petroleum jelly (Vaseline) in the nostrils every day can keep the nostrils from drying and cracking.

In some cases, local clotting agents such as thrombin and fibrin may be needed to prevent bleeding from starting again. The HTC team will be able to help with these treatments.

HEMATURIA

■ What is hematuria and how is it managed?

Hematuria refers to blood in the urine. It is caused by bleeding in the kidneys. A person with severe hemophilia will probably have an episode of hematuria at some time during his life. Hematuria generally disappears within a few days and permanent kidney damage does not occur.

If bleeding is mild, the urine will be pink and clotting factor therapy is not needed. The only treatment necessary is to make sure that your child drinks a lot and that there is a good output of urine. Since hematuria can be caused by conditions other than hemophilia, it is important to have your child checked by a doctor whenever there is blood in the urine.

If bleeding is persistent and the urine is dark red, treatment with clotting factor is needed. The HTC team will advise you on what dosage to give to treat hematuria. Factor therapy should be continued daily until bleeding stops. The antifibrinolytic agent tranexamic acid (Cyklokapron®) is **not** to be given if your child has hematuria.

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Management of Bleeds

Conclusion

This chapter has covered the management of common bleeding episodes including bruises, mouth bleeds, joint and muscle bleeds, nose bleeds and bleeding into the urine. It has also explained how to recognize life-threatening bleeds into the head, neck, chest and abdomen. Being able to recognize these types of bleeds as soon as they happen is key to successful treatment.

If you have any questions about how to treat a bleed, do not hesitate to contact the HTC team to discuss what you should do.

Finally, if your child is on a home infusion program and a bleed does not seem to be responding to treatment, you must take him to the hemophilia treatment centre to be examined by his doctor or another member of the care team.



Still a Mystery...

Many boys with hemophilia go through periods when they hardly bleed at all. Then, during other periods, they bleed often and for no reason. Is this caused by a small fluctuation in factor levels? Is it the result of another physiological cycle? Or is there a psychological explanation? Some people even say it is caused by the waxing and waning of the moon. It's still a mystery.