



INDIANA HEMOPHILIA & THROMBOSIS CENTER, INC.

helping patients thrive

Physical Therapy

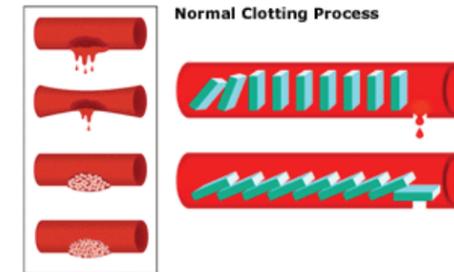
in Hemophilia Care

The Basics

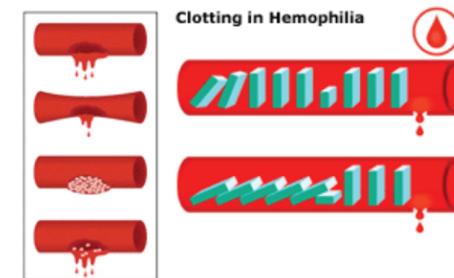
Hemophilia Basics

What is hemophilia?

Hemophilia is a rare disorder that disrupts normal blood clotting. This means that you bleed longer than a person without hemophilia, not faster. Normally, if you get a cut or hurt yourself, your body will protect itself by forming a scab or clot to stop the bleeding. Your body uses things called clotting factors to form a clot.



There are 12 clotting factors that you must have to form a stable blood clot. If you have hemophilia you are missing or have a low level of one of the clotting factors. Typically you are missing either clotting factor eight (factor VIII; hemophilia A) or clotting factor nine (factor IX; hemophilia B). A person with hemophilia is classified as having severe, moderate or mild deficiency based on the level of clotting protein found in the blood.



What are the common symptoms of severe, moderate and mild hemophilia?

Those with *severe* hemophilia often have frequent bleeding episodes if they do not receive preventative clotting factor replacement therapy, called prophylaxis. These individuals may experience bleeding in the joints, called a hemarthrosis, muscles and the mucous membranes of the nose and mouth. Bleeding episodes in those with severe hemophilia are often spontaneous and occur without a known cause. Persons with *moderate* hemophilia have less frequent bleeding episodes, typically due to an injury or surgery; however, some

spontaneous bleeding may occur in a person with a moderate deficiency. Those with *mild* hemophilia usually bleed infrequently and only with injury or during surgery.

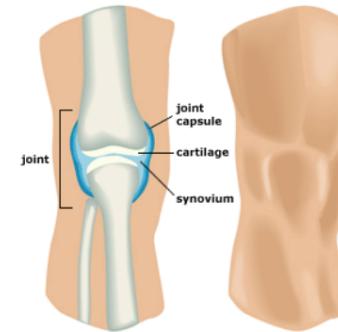
Hemophilia Severity	Clotting Factor Level in the Blood	Frequency of Spontaneous Bleeding
Severe	<1%	1 bleed/week
Moderate	1-5%	1-2 bleeds/year
Mild	>5-50%	occasional

The goal of this booklet is to describe what to expect if you have a joint or muscle bleed and the physical therapy measures that can be taken to assist in healing. Spontaneous joint and muscle bleeds are most common in individuals with severe hemophilia and occur less frequently in those with moderate hemophilia. Spontaneous joint and muscle bleeds are rare in individuals with mild hemophilia. Always consult your hemophilia treatment center physician before starting any exercise program.

Joint Bleeds

Hemophilia and Joint Bleeds

Anatomy of a Joint



A joint is where two or more bones come together (Figure 2). The ends of the bones are covered by a layer of cartilage that is important for cushioning and reducing friction during movement. Most joints are surrounded by a capsule that completely covers the joint. The inside of the capsule is lined by a thin layer of velvet-like tissue called synovium. The synovium plays an important role in maintaining a healthy joint. The synovium contains tiny blood vessels and produces a clear substance called synovial fluid. Synovial fluid provides nutrients to the joints, lubricates the joint structures, and removes waste from the joint area.

Figure 2.



What happens during a joint bleed?

When bleeding occurs in a joint, there is often a sensation of bubbling or tingling (Figure 3). This may occur before any outward signs of bleeding are noticeable. As the bleed continues, the joint capsule becomes swollen with blood (Figure 4). This causes swelling, pain or tenderness, heat or warmth, and loss of motion.

The blood that accumulates in the joint during a bleeding episode must be removed for the joint to function normally. The synovium, the velvet-like tissue surrounding the joint, must breakdown and absorb the blood within the joint. As the blood is broken down, several substances are released. These substances irritate and inflame the tissue and can damage the normal smooth, white cartilage which covers the ends of the bones.

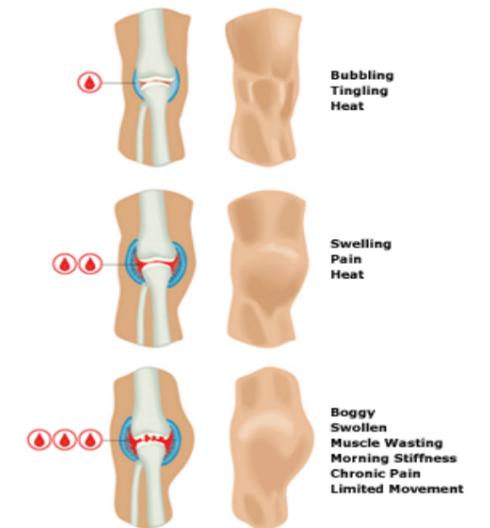


Figure 3.

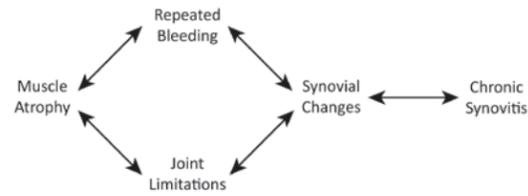


Figure 4. Right knee bleed.

Repeated episodes of bleeding can lead to a thickened synovium. A thick synovium is more easily traumatized which may lead to an increased risk of bleeding. Additionally, the ends of the bones become rough and bone spurs and arthritis develop over time.

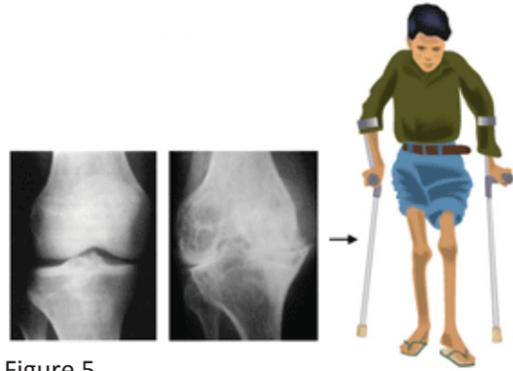
The joint becomes painful and the muscles around the joint decrease in size and strength. If appropriate treatment is not provided, a vicious cycle develops with repeated bleeding episodes which lead to muscle wasting, morning stiffness, chronic pain, limited movement, and arthritis.

Individuals with hemophilia who exercise regularly and maintain muscular health have fewer problems associated with spontaneous bleeding episodes. Through carefully chosen exercises and activity, individuals with hemophilia can maintain muscle strength and joint function and enjoy participation in sports.



What if I have repeated bleeding in the same joint?

Bleeding into muscles and joints is a common problem in individuals with severe hemophilia. Repeated bleeding into the same joint leads to what is known as a “target joint” and ultimately can lead to chronic synovitis and advanced degenerative arthritis (Figure 5). A target joint is formally defined as a joint that has three or more bleeding events over a six month period. Development of a target joint may lead to muscular and skeletal problems. Arthritis caused by repeated bleeding episodes can result in:



- Pain
- Decreased joint range of motion
- Muscle weakness
- Changes in the ability of the arms and legs to move normally
- An abnormal walking pattern
- Interference with activities of daily living such as personal hygiene, dressing, etc.

If you experience any of these symptoms it is important to contact your hemophilia treatment center for specific treatment instructions before engaging in the activities detailed below.

Figure 5.
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What should I do if I have a joint bleed?

The first 1-3 days following the beginning of a joint bleed is called the acute stage. The first goal during this stage is to stop the bleeding as soon as possible. This is achieved by infusing factor concentrate. Remember to infuse as early as possible to reduce the negative effects of continued bleeding in the joint; preferably within 3 hours of identification of a bleeding event. The acute stage lasts longer than 1-2 days, especially if factor concentrate infusions are not started immediately.

In addition to factor concentrate infusion, the **R.I.C.E.** method is often used to treat an acute joint bleed.

Rest the injured area to reduce the chance of re-bleeding. Also, reduce or stop using the injured area for at least 48 hours. If you have a leg injury, you may need to stay off of it completely. Crutches, splints or slings are often helpful if you need to remain active.

Ice the area using a cryocuff, cold pack, ice bag, or a plastic bag filled with crushed ice that has been wrapped in a towel. Ice limits blood flow to the injured area and can help reduce the pain associated with bleeding. Put an ice pack on the injured area for 10-20 minutes and then remove the ice for 10-20 minutes. Repeat this cycle every 1 to 2 hours.

Compression should be used to apply pressure to the bleeding ankle, knee or elbow. This may help reduce the swelling and provide support. You can apply pressure by using elastic wraps, special boots, air casts and splints. If using an elastic wrap, remember to start below the injured area and progress upward in a diagonal pattern as shown (Figure 6). Do not wrap the area too tightly. A good ‘rule of thumb’ is to make sure that you are able to

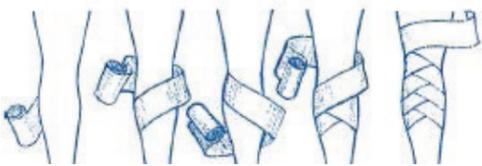


Figure 6.

insert one finger between the wrap and the affected limb. If an area is wrapped too tightly, numbness or

tingling, or a change in color or temperature may occur and the wrap should be removed immediately. Your hemophilia treatment center provider will instruct you in the proper wrapping technique.

Elevate the affected limb by keeping it *above the level of the heart*. Use a pillow.

If you experience any of these symptoms it is important to contact your hemophilia treatment center for specific treatment instructions before engaging in the activities detailed below.

When can I start using my joint again?

Once the bleeding in your joint has stopped, you may begin to move your joint. However, you should only move the joint in ways that do not cause pain or discomfort. You may still have some mild swelling, stiffness or limited range of motion in the joint. It is important **not to push through the pain** or overuse the joint as this can cause re-bleeding which leads to further joint damage. You may still need to wrap your joint for support and comfort during this time.

During this time you may also start to wean off assistive devices, such as crutches or a cane, depending on your level of pain. It is also important to remember that how well you recover depends on what caused your bleed and which tissues were injured. Your hemophilia treatment center physician may have you continue to infuse factor concentrate as your joint bleed resolves.

Why do I need physical therapy after a joint bleed?

Once the bleed has stopped, it is important to begin to re-gain your prior level of joint movement and activate the nearby muscles through physical therapy. Physical therapy will help reduce pain, swelling, and re-gain range of motion not always experienced with everyday activities. Physical therapy will also help strengthen the surrounding muscles which allows for more freedom of movement. If you have repeated bleeds in the same joint, you can progressively lose range of motion, muscle flexibility and develop muscle wasting.

Exercise for Joint Bleeds

Exercise Program for a Joint Bleed

Before describing the recommended exercise program for a joint bleed, a list of terms often used by physical therapists will be defined.

Definition of terms

Weights: Items with a certain heaviness lifted during an exercise. Weights can be items specifically designed for exercise, such as cuff weights or barbells, or they can be homemade. Items around the home that can be used to make weights include:

1. Cans of food held in the hand or placed in a satchel and lifted with the leg.
2. Sand or buckshot stitched into a bag or placed in double zip-lock bags.
3. Elastic resistance bands are elastic exercise bands that provide resistance to muscles through the range of motion. They vary in color which corresponds to resistance.

Repetitions or “reps”: The number of times that each exercise is done in a row.

Sets: Groups of repetitions, usually with rest periods between groupings.

Active Assistive Movement or Active Movement: Assisted movement of the joint by the patient and/or another person to achieve improved range of motion.

Isotonic or Active Movement: Movement of the joint by the patient through the joint’s range of motion.

Isometrics: Contraction of the surrounding muscle groups without actual movement of the joint.

Proprioception: The ability to sense the position, location, orientation, and movement of the body and its parts.

Resistive Exercise: Movement of the joint throughout the joint’s range of motion, with the addition of resistance band or weights to add resistance.

Range of Motion or ROM: The area through which a joint may normally be freely and painlessly moved.

Exercise Guidelines

The following are general exercise guidelines. This program is not tailored to meet every patient’s needs and thus you should consult with your hemophilia treatment center physician or physical therapist before beginning any exercise routine.

Once joint bleeding has resolved, swelling has decreased, and the joint moves with minimal-to-no pain, you may begin to exercise. These are basic guidelines and you may receive individual recommendations from your hemophilia treatment center.

1. Begin with *pain free* active movement. This type of exercise produces minimal stress on the joint and helps decrease the possibility of further bleeding episodes.
 - You may start with 1 set of 5 to 10 repetitions to determine your tolerance to the exercise. You can progress the exercise as tolerated to 3 repetitions of 10.

2. Isometric muscle contractions may begin at this time. It is important to address all muscles surrounding the joint. Initially you may be unable to do a full muscle contraction and thus starting with a light muscle contraction may be necessary.
 - The isometric exercise can be held to a count of 3-5 seconds to begin.
 - You may start with 1 set of 5 to 10 to check your tolerance to the exercise. You can progress the exercise as tolerated to 3 sets of 10.
3. Once full *pain free* range of motion and muscle flexibility has been achieved, you progress to resistance exercises to increase muscle strength.
 - You may start with light weights such as 1-2 pounds in sets of 5 to 10 to check your tolerance to the exercise. You can progress the exercise as tolerated to 3 sets of 10.
4. Proprioception exercise will only be initiated once full weight bearing and range of motion is achieved. These specific exercises will be provided by your hemophilia treatment center physical therapist.
5. Depending on the recovery process, outpatient physical therapy may be recommended. The hemophilia treatment center physical therapist may provide outpatient therapy. However, if you do not live close to the hemophilia treatment center, they will assist you to identify an appropriate therapy facility. Additionally, they will provide education to your assigned therapist regarding hemophilia.

Muscle Bleeds

Hemophilia and Muscle Bleeds

Anatomy of a Muscle

The skeletal muscles of your body are made up of many fibers. These fibers shorten or lengthen as needed to allow movement. The muscle tissue contains many blood vessels which supply oxygen and nutrients to the muscle. Muscles are grouped into different sections within our arms and legs. These sections contain muscles, nerves and blood vessels which are surrounded by a tough, non-stretchy connective tissue known as fascia.

What happens during a muscle bleed?

Most muscle bleeds are a result of injury or a muscle strain. A muscle strain is commonly referred to as “pulled muscle”. It can occur during vigorous activity, sports participation or lifting. A strained muscle can result in tearing of the muscle fibers. This causes bleeding into the muscle (Figure 7). Direct injury to a muscle, known as a contusion, can damage the underlying muscle tissue and cause bleeding. The accumulation of blood in a muscle is called a hematoma. Frequent bleeding into a muscle can lead to muscle deterioration, called muscle atrophy. This can lead to your muscle becoming less flexible and may cause permanent shortening of the muscle, called a contracture, which leads to limited joint movement.



Figure 7. (Left) A calf bleed; (Right) A thigh bleed.

As blood collects in the muscle, it will cause the muscle to swell. Larger muscles in the thigh or buttocks may take longer to show signs of swelling. Common signs of a muscle bleed may include:

- Muscle pain, swelling, warmth
- Muscle spasm
- Limited ability to move the affected area
- Increased pain with muscle movement, stretching, or weight bearing
- Tissue over the muscle may become tight and sometimes appear shiny
- An accumulation of blood under the skin
- A bruise may be present
- Interruption of normal nerve or blood flow function

If the muscle continues to bleed, a serious condition known as compartment syndrome may develop. Because muscles are surrounded by a non-stretchy tissue called the fascia, when blood accumulates it can put pressure on the surrounding nerves and blood vessels. The increased pressure can press against the nerves and stop blood flow. This can lead to permanent nerve and muscle damage. Common sites for a compartment syndrome include the forearm muscles, calf muscles, and thigh muscles; however, any muscle compartment can be affected.

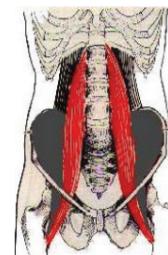


Figure 8.

Anatomy of the Iliopsoas Muscle

The iliopsoas muscle, *shown in red* in Figure 8, is actually made of three muscles: the iliacus muscle and the psoas major and minor muscles. Although these muscles start at different places on the spine and pelvis, they come together at the thigh and are often referred to as one muscle. This muscle is responsible for bending the hips and is used to run, walk and stand. It can also move your lower back.

What is an iliopsoas bleed?

Iliopsoas bleeds are relatively common, but may be difficult to identify, and recovery can be slow. This muscle is located deep inside the abdomen and there are usually no outward signs of swelling or bruising. This is a large muscle and thus you may experience prolonged bleeding before experiencing symptoms. Common signs of an iliopsoas bleed include:

- Pain in the lower groin, lower abdomen and/or lower spine area
- Decreased sensation or tingling on the front of the thigh
- Pain with weight bearing
- Pain when walking
- Difficulty using the hip and keeping the hip in a flexed position of comfort
- An arched back when standing or lying down
- Difficulty lying flat when the legs are extended straight

If you experience any of the symptoms commonly associated with a muscle bleed, it is important to contact your hemophilia treatment center for specific treatment instructions before engaging in the activities detailed below.

What should I do if I have a muscle bleed?

The most common sites of a muscle bleed include: the hamstring (back of the thigh), buttocks (in children), calf, biceps (upper arm), and forearm. Just as with a joint bleed, the first 1-3 days following the onset of a joint bleed is considered the acute stage. The primary goal during this stage is to stop the bleeding as soon as possible by infusing clotting factor concentrate. It is important to infuse immediately, optimally within 3 hours of identification of the bleed, to reduce the adverse effects of prolonged bleeding into the muscle. The acute stage may last longer than 1-3 days, especially if clotting factor concentrate infusions are started late. You should always contact your hemophilia treatment center for specific instructions.

In addition to clotting factor concentrate infusion, using the **R.I.C.E.** method can help control swelling, pain and protect the injured muscle tissues.

Rest the injured area to reduce the chance of re-bleeding. Also, reduce or stop using the injured area for at least 48 hours. If you have a leg injury, you may need to stay off of it completely. Crutches, splints or slings are often helpful if you need to remain active.

Ice the area using a cryocuff, cold pack, ice bag, or a plastic bag filled with crushed ice that has been wrapped in a towel. Ice limits blood flow to the injured area and can help reduce the pain associated with bleeding. Put an ice pack on the injured area for 10-20 minutes and then remove the ice for 10-20 minutes. Repeat this cycle every 1 to 2 hours.

Compression may not always be used during an acute muscle bleed as the added pressure may not be tolerated.

Elevate the affected limb by keeping it *above the level of the heart*. Use a pillow.

When can I start using my muscle again?

Your hemophilia treatment center physician may have you continue to infuse clotting factor concentrate as your muscle bleed resolves.

Muscle bleeds tend to resolve more slowly than joint bleeds. Once the area is no longer tense or painful, you can begin to move your muscle in pain-free movement. You should avoid aggressive stretching as this may cause re-bleeding. You may still have swelling, stiffness or limited range of motion and it is important **not to**

push through the pain or over-use the muscle as this can also lead to re-bleeding. Wrapping the muscle for support may provide comfort and support during this time. Additionally, you may start to wean off assistive devices such as crutches/cane as determined by your level of pain.

It is important to recognize that depending on the cause of your muscle bleed, there may be underlying muscle and skeletal tissue injury which can influence your recovery.

Why do I need physical therapy after a muscle bleed?

Physical therapy will help reduce pain and swelling and will assist you to re-gain muscular function, range of motion. This will help you return to your prior level of function and mobility.

During a muscle bleed you will have reduced use of the muscle due to pain, loss of range of motion and muscle spasm. Once the bleed has resolved, it is important to begin to re-gain your full motion and activate the muscle. Physical therapy exercises will help reduce the swelling, improve your muscle flexibility and re-gain your strength in the affected muscle. The physical therapist will provide you with exercises that will ensure normal muscle balance.

Exercise for Muscle Bleeds

Exercise Program for a Muscle Bleed

Before describing the recommended exercise program for a muscle bleed, a list of terms often used by physical therapists will be defined.

Definition of terms

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Once joint bleeding has resolved, swelling has decreased, and your pain has decreased, you may initiate movement by exercising the area.

1. Begin with *pain free* active movement. This type of exercise will begin to use the muscle and move the joint.
 - You may start with 1 set of 5 to 10 repetitions to determine your tolerance to the exercise. You can progress the exercise as tolerated to 3 repetitions of 10.

2. Isometric muscle contractions may begin at this time. It is important to address the muscle on the opposite, surrounding joint as well. Initially you may be unable to do a full muscle contraction and thus starting with a light muscle contraction may be necessary.
 - The isometric exercise can be held to a count of 3-5 seconds to begin.
 - You may start with 1 set of 5 to 10 to check your tolerance to the exercise. You can progress the exercise as tolerated to 3 sets of 10.
3. Once full *pain free* range of motion and muscle flexibility has been achieved, you progress to resistance exercises to increase muscle strength.
 - You may start with light weights such as 1-2 pounds in sets of 5 to 10 to check your tolerance to the exercise. You can progress the exercise as tolerated to 3 sets of 10.
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5. Depending on the recovery process, outpatient physical therapy may be recommended. The hemophilia treatment center physical therapist may provide outpatient therapy. However, if you do not live close to the hemophilia treatment center, they will assist you to identify an appropriate therapy facility. Additionally, they will provide education to your assigned therapist regarding hemophilia.

Additional Therapy & Assistive Devices

Additional Therapy Measures and the Use of Assistive Devices

The use of additional modalities to facilitate an exercise program is also recommended. Although some of these treatments require a visit to a physical therapist, most can be learned by the patient and incorporated into a home exercise program.

- **ASSISTIVE DEVICES:** Assistive devices, such as canes and crutches may be necessary to help rest a joint or muscle, especially during an acute bleeding episode. Assistive devices should be used only when necessary. If used for a prolonged period of time, assistive devices can increase the chance of causing bleeds in the upper extremities, especially the elbows.
- **ELECTRICAL STIMULATION:** Electrical stimulation can be used to stimulate a muscle contraction for muscle strengthening. A process called transcutaneous electric nerve stimulation, or TENS, may be used for pain control.
- **HEAT:** Heat can be used to help relieve pain and promote relaxation. DO NOT use heat during the acute phase of a bleeding episode.
- **ICE:** The use of ice has been mentioned in the treatment of acute joint and muscle bleeds. Ice may also be effective before or after an exercise session to reduce inflammation.
- **MASSAGE and RELAXATION:** Massage, relaxation and visual imagery can help relax tense muscles. This may alleviate stress and decrease pain related to bleeding episodes.
- **SPLINTS and BRACES:** Elastic, neoprene, splints and arch supports may be used to support a joint and some adjacent muscles. These devices help protect the joint; however, some can restrict joint movement. Restricted joint movement may result in muscle wasting and weakness and thus an active strengthening program is necessary to maintain normal strength.

The ultimate objective in physical therapy and hemophilia care is to restore each individual to their highest level of musculoskeletal performance and activity after a bleeding episode. To do this, it is important to incorporate regular exercise or sports into your everyday life. Specific sports such as swimming, weight lifting, golf, tennis, bicycling, and brisk walking are recommended for those with hemophilia to maintain muscle strength and cardiovascular fitness. A healthier body is linked to a healthier mind and this improves your quality of life.

Procedures

Exercises

