

FACTOR XIII
DEFICIENCY

AN INHERITED
BLEEDING DISORDER

AN INFORMATION
BOOKLET

Acknowledgements

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Introduction

Factor XIII Deficiency is a very rare disease. It is not well known, even among health professionals. There is very little written information available to people with Factor XIII Deficiency and their families. The purpose of this booklet is to describe the disease and its treatment. We hope that it will permit those affected to better understand their disorder and reduce its impact on their lives.

How Factor XIII Deficiency is Inherited

Factor XIII Deficiency is an inherited bleeding disorder. It is passed on from parent to child at the time of conception. The bleeding problem is caused by an abnormal gene.

Each cell of the body contains structures called *chromosomes*. A chromosome is a long chain of chemicals known as DNA. This DNA is arranged into about 200,000 units called genes. These genes determine such things as the colour of a person's eyes. In the case of Factor XIII Deficiency, one of these genes has a defect.

The defective gene in Factor XIII Deficiency is on a chromosome that does not decide the sex of the child. This means that Factor XIII Deficiency can affect females as well as males. In this way, it is unlike other bleeding disorders such as Factor VIII Deficiency (hemophilia A), in which the defective gene is sex-linked—and therefore only males are affected.

A *carrier* is a person who carries the defective gene but is not affected by the disease. In order for a person to inherit Factor XIII Deficiency, both parents must be carriers. In such a case, the baby inherits two defective Factor XIII genes, one from the mother and the other from the father.

If a person inherits the defective gene from only one of the parents, he/she will be a carrier. His/her Factor XIII level will be below normal, but there will be no signs of the disease.

The three illustrations below show how Factor XIII Deficiency can be passed on.

Figure 1 shows what can happen when a carrier of Factor XIII Deficiency has children with another carrier. There is a 1-in-4 chance that a child will have severe Factor XIII Deficiency, a 1-in-2 chance that a child will be a carrier and a 1-in-4 chance that a child will be normal.

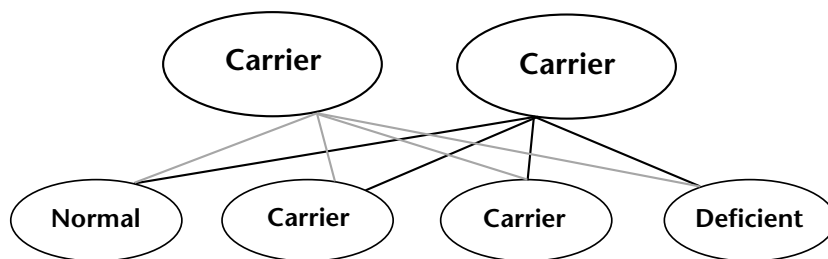


Figure 1

Figure 2 shows what can happen when someone with severe Factor XIII Deficiency has children with a non-carrier. All the children will be carriers, but none of them will have the disease.

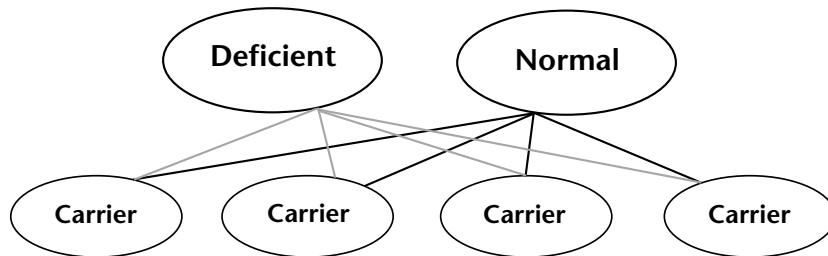


Figure 2

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Figure 3 shows what can happen when someone with severe Factor XIII Deficiency has children with a carrier. There is a 1-in-2 chance that a child will be a carrier. There is also a 1-in-2 chance that a child will have severe Factor XIII Deficiency.

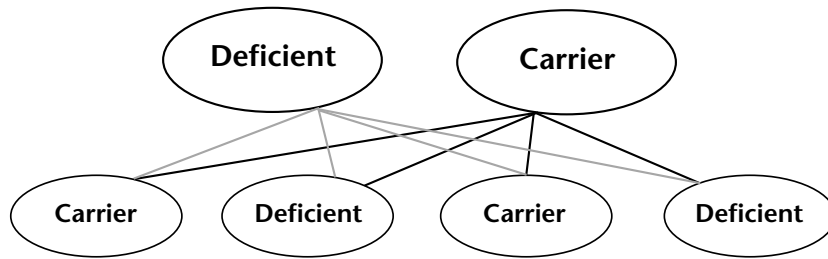


Figure 3

Since Factor XIII Deficiency runs in families, and since the defective gene is so rare, there is often a history of consanguinity (people marrying close relatives) in cases of this disease.

The Cause of Bleeding in Factor XIII Deficiency

Blood is carried throughout the body in a network of blood vessels. When tissues are injured, damage to a blood vessel may result in leakage of blood through holes in a vessel wall. The vessels can break near the surface, as in the case of a cut. Or they can break deep inside the body, causing a bruise or an internal *hemorrhage*.

Clotting, or coagulation, is a complex process that makes it possible to stop injured blood vessels from bleeding. As soon as a blood vessel wall breaks, the components responsible for coagulation come together to form a plug at the break. There are several steps involved in forming this plug.

- *Blood platelets*, which are very tiny cell fragments, are the first to arrive at the break. They clump together and stick to the wall of the damaged vessel.
- These platelets then emit chemical signals calling for help from other platelets and from clotting factors.
- The clotting factors, which are tiny plasma proteins, link to form a chain, called *fibrin*. The strands of fibrin join together to weave a mesh around the platelets. This prevents the platelets from drifting back into the blood stream.
- Then, Factor XIII plays a special role in stabilizing the fibrin mesh by solidifying the individual fibres. Without Factor XIII, the fibrin strands are fragile and individual fibres break away. And when this happens, bleeding begins again.

Factor XIII is a protein made up of two parts. One is produced in the bone marrow; the other in the liver. A liver transplant can cure a hemophiliac with Factor VIII or Factor IX Deficiency, diseases that are much more common than Factor XIII Deficiency. This is because Factor VIII and Factor IX proteins are produced entirely in the liver. In Factor XIII Deficiency, because some of the proteins are produced in the bone marrow, a liver transplant is not a cure.

■ Diagnosis

When a patient shows signs of unusual bleeding, a family doctor will often order several blood tests to measure coagulation. These tests measure the time it takes for a clot to form. The clot is a kind of mesh composed of strands of fibrin. In many kinds of bleeding disorders, a clot will take a long time to form. In other words, the clotting time will be longer than normal.

However, with Factor XIII Deficiency, the clot formation takes the usual time – the clotting time is normal. These tests are therefore misleading.

As mentioned above, the role of Factor XIII is to solidify the fibrin strands that make up the clot. When Factor XIII is absent, and the chains are weak, the strands break easily. The clot is unstable, it breaks down and further bleeding occurs. This can happen hours or even days after the injury first occurs. This is why clotting time results are normal in Factor XIII Deficiency, even though there is a very real clotting problem.

To diagnose Factor XIII Deficiency accurately, lab technicians perform a test of the *quality* of Factor XIII in the blood sample. This test does not measure the exact *quantity* of Factor XIII, since even a very small amount (about 2% of normal) is enough to stabilize the clot and prevent bleeds.

If Factor XIII Deficiency is found, a test of the exact amount of Factor XIII in the blood is done. The result of this quantitative test shows the severity of the Factor XIII Deficiency. People with this disease have less than 2% of the normal amount of Factor XIII. The closer the level is to 0%, the more severe the disease.

The quantitative test can also be used to find out if a person is a carrier. In people who neither have Factor XIII Deficiency nor carry it, the level of Factor XIII is about 100% of normal. In carriers, it is about 50%, and in those with Factor XIII Deficiency, it is less than 2%.

Incidence

Factor XIII Deficiency is an extremely rare disorder. It affects only one in several million people. Fewer than ten Canadians have been diagnosed with the disease. To date, only about 200 cases have been reported worldwide. It is found in people of all races and ethnic origins.

Symptoms

The most common symptoms of Factor XIII Deficiency are:

- persistent bleeding from the umbilical stump a few days after it falls off. This is seen in about 80% of cases of Factor XIII Deficiency.
- bleeding in soft tissues. This takes the form of bruises (accumulation of blood under the skin).

The following symptoms are less common. About 30% of patients with severe Factor XIII Deficiency experience the following types of bleeding:

- central nervous system bleeding (in the brain or the spinal cord). This can happen with or without injury.
- mouth bleeds.
- intramuscular bleeds (accumulation of blood in a muscle). This can occur after strenuous exercise even if there has been no injury.
- bleeding from a laceration (cut).

Bleeding into joints (called hemarthrosis) is quite rare. More common is bleeding around the joints (periarticular bleeding).

Bleeding into soft tissues and muscles may also lead to pseudotumours. Large swellings form due to the accumulation of blood. These are caused by repeated or long-term bleeding. Pseudotumours can be found in the thighs, calves, buttocks and other fleshy areas.

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Bleeding during surgery is not excessive. However, further bleeding may occur hours or days after the operation.

Healing of wounds is more difficult for people with Factor XIII Deficiency. Factor XIII plays a role in cementing a clot, as well as in tissue healing and strength. A severe deficiency in Factor XIII hinders the formation of scar tissue.

Bleeds Associated with Factor XIII Deficiency

Frequent

- Umbilical stump bleeding after birth
- Superficial bruises
- Bleeding into soft tissues
- Central nervous system bleeding

Less Frequent

- Bleeding from the gums, into muscles and joints; cuts, scrapes
- Bleeding in and around the abdomen
- Bleeding after surgery

Rare

- Nose bleeds
- Genital and kidney bleeds
- Eye bleeds
- Gastrointestinal bleeds
- Bleeding in the spleen, lungs or ears

Treatment and Prevention of Bleeding

All that is required to control and prevent bleeding is to increase slightly the amount of Factor XIII in the blood.

Because of the high risk of bleeding in the brain (intracranial hemorrhage), doctors recommend preventive treatment for a person with Factor XIII Deficiency. Preventive treatment means regular infusions of Factor XIII, even if there is no sign of bleeding. This therapy is called *prophylaxis*.

Blood products used in the treatment of Factor XIII Deficiency have changed a lot over the years. Plasma and cryoprecipitate, good sources of Factor XIII, were used in the past. However, they are no longer recommended. The reasons are:

- There is a tiny risk of viral transmission. We know today that HIV and hepatitis B and C (which cause liver disease) can be transmitted through blood products. As a result, blood donors are carefully selected. They are questioned to be sure they are not at high risk of carrying a virus. Their donations are tested to detect signs of HIV and hepatitis B and C viruses. Today, the risk of infection from a blood transfusion is extremely small. However, a tiny risk remains. A blood donor who has been very recently infected by HIV or hepatitis B or C viruses may not yet have developed the telltale signs that permit his/her donation to be rejected.
- Plasma and cryoprecipitate can cause severe allergic reactions. This is because they contain many different substances in addition to Factor XIII.
- The quantity of Factor XIII in a bag of plasma differs from one bag to the next, even when the amount of plasma is the same. This means it is impossible to know exactly how much Factor XIII is being administered.
- There is very little Factor XIII in a bag of plasma. To control or prevent bleeding, a large volume of plasma must be infused slowly over several hours. This can overload the heart.

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Today, to treat Factor XIII Deficiency, a Factor XIII *concentrate* is used instead of plasma or cryoprecipitate. It comes in the form of a freeze-dried powder stored in a small vial. When it is time for a treatment, sterile water is added to the vial and the powder is dissolved. It is then infused into a vein. The treatment only takes a few minutes.

This concentrate is made from the pooled plasma of thousands of blood donors. Here are the reasons why Factor XIII is a better choice than plasma or cryoprecipitate.

- Factor XIII concentrate undergoes a viral inactivation process that kills viruses that may be present in the blood. Before being placed on the market, each batch is tested to ensure it is virus-free. Each batch, or lot, of Factor XIII concentrate is made from plasma from a single group of donors.
- Because of the greater purity of concentrate, allergic reactions are extremely rare.
- Factor XIII concentrate comes in a small vial and is measured in units. The exact quantity contained in the vial is clearly marked. It is easy to determine the exact dosage of Factor XIII administered.
- The product is highly concentrated. The amount necessary to treat or prevent bleeding is diluted in less than 20 millilitres of sterile water. Therefore, there is little danger of overloading the heart. The treatment can be administered in just a few minutes.

Like other proteins in our bodies, Factor XIII has its own life cycle. It forms, lives and dies on a fixed timetable. In a person with Factor XIII Deficiency, Factor XIII is not automatically renewed, since he/she lacks the gene required for its fabrication. The Factor XIII protein contained in the concentrate is identical to that manufactured by the body. It, too, has a limited life span. The term used to measure how long the Factor XIII survives in the body is *half-life*. The half-life is the amount of time it takes for half the infused Factor XIII to be removed from the bloodstream.

To determine how long to wait between doses of Factor XIII, its half-life must be taken into account. Factor XIII has a half-life of 8 to 10 days. In other words, half of the Factor XIII administered will have disappeared 8 to 10 days after its infusion.

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In order to control or prevent bleeding, the level of Factor XIII must never be less than 1% of normal. The following information explains how to calculate the dose and frequency of Factor XIII infusions so that a person always has enough clotting factor in his/her blood.

The infusion of one unit of Factor XIII per kilogram of body weight increases Factor XIII levels by about 1.5%. For example, if a person weighing 50 kg receives an infusion of 10 units of Factor XIII per kilogram of body weight (that is, 10 units/kg X 50 kg = 500 units), then the level of Factor XIII rises by 10 times 1.5%, or 15%. (Remember that 1 unit/kg increases Factor XIII content of the blood by 1.5%.) At 15% of normal, this person is well protected against bleeding.

After 10 days, the level of Factor XIII in his/her blood drops by about half (15% divided by two) to about 7.5%. This is because the half-life of Factor XIII is about 10 days, and half the Factor XIII is gone. The patient continues to be protected.

After another 10 days, that is 20 days after the infusion, the person's Factor XIII level again goes down by a half (7.5% divided by two) to about 3.75%. This is still a satisfactory level of protection.

After another 10 days (30 days since the treatment), the level of Factor XIII drops to about 1.9% (that is, 3.75% divided by two). The patient now needs a new infusion of concentrate to raise his or her Factor XIII to a safe level.

In other words, a monthly treatment with about 10 to 20 units per kilogram of body weight, that is, 500 to 1000 units for an adult, is generally enough to ensure that there will be no abnormal bleeding.

However, before deciding on a treatment program for a patient, a Factor XIII half-life test is recommended. This involves infusing Factor XIII concentrate, and then measuring Factor XIII levels at regular intervals over a period of a month. These tests will tell doctors what amount of product is needed so that the Factor XIII levels never drop below 1%. The dosage and treatment frequency can then be adjusted to each individual.

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The Factor XIII concentrate currently on the market comes in vials of 250 or 1250 units. The dose of Factor XIII is rounded off so that the entire contents of the vial are administered. This product is extremely expensive, and should not be wasted. The different vial sizes allow for infusion of the correct dose. The entire vial should always be infused as a higher than prescribed dose is harmless.

A person with severe Factor XIII Deficiency should learn how to self-infuse Factor XIII concentrate at home. Self-infusion has many advantages.

- It allows the person considerable independence.
- It removes the inconvenience of having to go to a healthcare centre every month.
- If a bleed occurs, the person has access to rapid, effective treatment without having to go to a healthcare centre. Factor XIII Deficiency is a very rare disease. Few health professionals know much about it. At a hospital ER, precious minutes could be wasted studying the patient's records, trying to reach the attending physician or running through long and tedious medical examinations. This may result in an unnecessary delay in the infusion of Factor XIII concentrate while a potentially dangerous bleed continues.

As soon as a person is informed of his/her diagnosis of severe Factor XIII Deficiency, he/she should receive an ID card naming the disease, describing the appropriate treatment, and giving the name of the Hemophilia Treatment Centre and the telephone numbers of health professionals on the healthcare team. In the case of an accident or an emergency surgery, this card will be very helpful for the patient and the caregiving team.

A person with Factor XIII Deficiency who lives far from his/her treatment centre or who does not want to learn how to self-infuse, should have a summary of his/her medical records sent to the hospital closest to his/her home. The information should include appropriate measures to take in case of bleeding. A permanent prescription for Factor XIII should be given to the nurses so they can treat the patient on his/her arrival at the hospital, even before he/she is examined by a doctor.

Recognizing Bleeding

People with Factor XIII Deficiency are considered to be hemophiliacs. As in other types of hemophilia, they bleed abnormally due to a clotting factor deficiency. However, the nature, frequency and treatment of bleeding in Factor XIII Deficiency are very different from those in other types of hemophilia.

- Umbilical and intracranial bleeds are much more frequent in Factor XIII Deficiency than in hemophilia A or B (Factor VIII and IX Deficiencies).
- Joint and muscle bleeds are much less frequent in Factor XIII Deficiency than in Factor VIII or Factor IX Deficiency.
- Preventive therapy (prophylaxis) is much more effective in this disorder because of the longer half-life of Factor XIII. The half-life of Factor XIII is 8 to 10 days, that of Factor VIII about 12 hours and that of Factor IX about 18 hours. The result is that there is almost no bleeding once Factor XIII concentrate is administered in adequate doses at suitable intervals.

It is strongly recommended that patients with Factor XIII Deficiency learn to recognize the signs and symptoms of potentially life-threatening bleeds, so they can respond in a timely and appropriate manner. The following information describes the main types of bleeding that can occur in a person with a bleeding disorder.

Bleeds which occur in the head, neck, thorax (chest) or abdomen (stomach) can be life-threatening and require immediate medical attention. It is important to realize that these bleeds can occur following an injury or spontaneously (without an injury).

HEAD

The brain, protected by the skull, controls all functions in the body required to keep a person alive. A bleed to the brain is extremely serious.

Signs and Symptoms

- Headache
- Problems with vision (eyesight)
- Nausea and vomiting
- Personality changes
- Sleepiness
- Loss of balance
- Loss of fine motor coordination (clumsiness)
- Fainting
- Convulsions

Any of these symptoms should alert a person to take action and to get immediate treatment. If any of these symptoms occur, a physician should be contacted immediately.

NECK

The tissues in the neck, mouth and throat are extremely *vascular*. This means that they contain many veins and arteries that carry blood. A small injury or infection can cause an accumulation of blood in these tissues. As the tissues slowly fill with blood, they increase pressure on the respiratory tract. This makes breathing difficult and can even block the airway completely.

Signs and Symptoms

- Pain in the neck or throat
- Swelling
- Difficulty swallowing
- Difficulty breathing

THORAX (chest)

The chest cavity (thorax) contains the lungs, heart and many large blood vessels. Bleeding into the lung tissue traps blood inside the air sacs which normally hold oxygen. This makes breathing difficult.

Signs and Symptoms

- Pain in the chest
- Difficulty breathing
- Coughing or spitting up blood

ABDOMEN (belly)

The stomach, spleen, liver and intestines are but four of the organs found in this cavity. An injury in this area can cause massive bleeding in the organs themselves or from a large blood vessel. Failure to treat this type of bleed could be fatal.

Signs and Symptoms

- Abdominal pain or lower back pain
- Nausea and vomiting
- Presence of blood in the urine
- Presence of blood in the stools or black stools

If any of these symptoms occur, the patient must seek immediate medical treatment.

There are other types of bleeding which require medical treatment but which are not as serious. These are discussed below.

Soft Tissue Bleeding

The signs and symptoms of soft tissue bleeds are:

- **Redness at the injury site** – A measuring tape should be used to measure the site. If a measuring tape is not available, check the site every hour to judge if the area of redness is getting bigger.
- **Increasing size of a bruise** – A pen should be used to outline the outside of the bruise to see if the bleed is getting bigger, smaller, or if it is stable.
- **Pain** – The patient should take note whether or not the pain gets worse with time, especially if he/she can pinpoint the exact site of the pain.

Joint Bleeding

The signs and symptoms of joint bleeds are:

- Pain during normal use of the joint or even while resting, especially if there is no sign of bruising.
- Swelling and warmth, with or without bruising.
- Decrease in the normal movement of a joint.
- Protecting a joint – For example, a child who walks normally will suddenly develop a limp, due to an ankle bleed. A right-handed child will start using his left arm to grasp objects, due to an elbow bleed.
- Fussing or crying during movement of a joint – This could suggest pain due to a joint bleed, especially in infants. Parents must learn to examine the joints and assess joint movement. This is especially important for the knees, ankles, and elbows.

Basic First Aid to Treat Bleeding

This section describes how to treat minor and moderate bleeds in joints or in soft tissues.

Rest, ice, compression and elevation are four ways of lessening the pain and discomfort of a bleed.

Rest - Rest a limb by using crutches or a wheelchair. Minimize walking as much as possible. Rest an arm by using a scarf or a sling to support the limb.

Ice - Apply ice to the injured site. Use an ice-pack or a bag of frozen vegetables wrapped in a damp towel. Never apply the ice directly to the skin. Apply the ice for about 15 minutes every 2 hours.

Compression - Wrap an injured joint in an elastic bandage using a figure-eight pattern. Watch for signs of numbness, cold, sharp pain, or a change of colour in the finger or toes. These are signs that the circulation has been cut off. If any of these signs occur, remove the bandage and reapply it with less tension.

Elevation - Lift the affected limb above the heart to reduce swelling. This will also improve blood circulation.

The Hemophilia Treatment Centre team will provide support during these bleeding episodes.

A major bleed into a muscle can cause permanent damage to the affected limb. The muscles and arteries can be compressed by the accumulation of blood. If you think you, or your child, might have a muscle bleed, contact your treatment centre immediately. In addition, watch carefully for the signs described above with regards to compression with an elastic bandage.

Issues Concerning Reproduction

Most pregnancies in women with severe Factor XIII Deficiency end in miscarriage (spontaneous abortion), unless the patient receives Factor XIII concentrate regularly during her pregnancy. Among women who are carriers of Factor XIII Deficiency (but do not have the disorder), the rate of miscarriage is higher than in the general population.

The reasons why Factor XIII is necessary for the continuation of pregnancy are not well known. The normal placenta has a high Factor XIII content. This is perhaps necessary to prevent uterine or placental bleeding or to promote adherence and growth of the placenta.

The duration and intensity of menstruation are the same in women with Factor XIII Deficiency as in women in the general population. Thus, this disorder has no effect on the menstrual cycle. However, as stated above, it does affect the possibility of carrying a pregnancy to term.

In men, lower sperm counts (oligospermia) and infertility are associated with Factor XIII Deficiency.

Lifestyle

Since prophylactic therapy offers excellent protection to patients with Factor XIII Deficiency, lifestyle recommendations are, in general, the same as for anyone in good health.

- Follow a regular program of physical activity suited to your taste and capability. Use high quality sports equipment appropriate for your sport (helmet, elbow pads, kneepads, proper shoes, etc.).
- Eat a well-balanced diet; maintain a normal weight.
- Get enough sleep.
- Drink alcohol in moderation only.
- Don't smoke.

Here are a few other recommendations specifically for patients with Factor XIII Deficiency.

- Maintain good dental hygiene and see your dentist regularly. If a tooth has to be extracted, time the visit to your dentist to coincide with your preventive dose of Factor XIII. If necessary, treat with Factor XIII concentrate before the visit. Your treatment centre can recommend a dentist who is thoroughly familiar with hemophilia.
- When surgery is planned, contact your treatment centre to plan appropriate preventive therapy. In case of emergency surgery, the physician on duty can contact a member of the treatment centre team at any time by calling the number on your patient ID card.
- **Never take aspirin.** Aspirin increases the risk of bleeding. It affects the way platelets clump together to plug a broken blood vessel. Always consult the healthcare team before taking any new medication, any medicinal-herb-based supplement or any vitamins sold over the counter.
- Wear a MedicAlert-type bracelet or chain at all times on which the type of coagulation problem you have is engraved.

- Always keep in mind the signs and symptoms of life-threatening bleeding—particularly those of intracranial bleeding, which is common in Factor XIII Deficiency.
- If you are planning a trip, let your treatment centre know. You should have a letter from your doctor describing the medical supplies you have to take with you (syringes, needles and medication) so you can cross borders without any problems. Make sure you have enough Factor XIII for the duration of your trip. Ask for the addresses of hemophilia treatment centres in the area you are visiting.
- Always stay in touch with your hemophilia treatment centre.

Vaccination

Children should be vaccinated on a timetable set by their pediatrician or family doctor. However, anyone receiving clotting factor concentrates of human origin should be vaccinated against hepatitis A and hepatitis B.

Hepatitis A, on very rare occasions, has been transmitted by modern factor concentrates. Hepatitis A is an infection caused by a virus. It affects the liver. It is not the same virus as the one that causes hepatitis B. Some people have relatively severe symptoms; others don't even know they have it. An infected person can transmit the disease to others. Hepatitis A is rarely fatal. Only the elderly and those with chronic liver disease (cirrhosis or hepatitis C) run a risk of death from hepatitis A. The symptoms gradually disappear after a few weeks. Once cured, the patient is protected against hepatitis A for life and is no longer a carrier of the virus.

Hepatitis A is found in the stools of anyone who is infected. It can be spread by:

- drinking contaminated water or eating food that has been in contact with an infected person
- sexual relations with an infected person
- contact with infected blood.

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Hepatitis B used to be transmitted by factor concentrates. Today's concentrates are considered safe. However, a person with a bleeding disorder has a greater chance of needing a blood transfusion. Blood transfusions can still, on very rare occasions, transmit hepatitis B. For this reason, vaccination is recommended.

Hepatitis B is a disease caused by a virus that attacks the liver. The liver helps to digest food and clean the blood. Sometimes, people with hepatitis B do not feel sick at all, but they can still transmit the disease. In other cases, hepatitis B makes people very sick. It can cause serious damage to the liver and an infection that lasts a very long time. There is no treatment that is completely effective in curing hepatitis B.

Hepatitis B is transmitted by contact with body fluids:

- blood
- breast milk
- sperm
- vaginal secretions

There is a combination vaccine for protection against hepatitis A and hepatitis B, called Twinnix. It is administered in three doses. The booster is given one month after the first dose and a third dose is given six months later.

The two vaccines can also be administered separately. The vaccine against hepatitis B is also administered in three doses, with boosters one month and six months after the initial dose. The hepatitis A vaccine is administered in two doses, the second dose about six to 12 months after the first.

The Comprehensive Care Team

As the name suggests, a hemophilia comprehensive care team provides most of the medical services required by a child or adult with an inherited bleeding disorder. The team is composed of several professionals, including:

- a medical director
- a nurse coordinator
- a physiotherapist and
- a social worker.

The team works closely with other specialists – a surgeon, an orthopedist, a rheumatologist, a dentist, a geneticist and a psychiatrist, among others. The purpose of this multidisciplinary team is to ensure the well being of the patient and, in the case of a child, of the parents as well.

Summary

Factor XIII Deficiency is an inherited blood clotting disorder that affects women as well as men. In severe Factor XIII Deficiency, life-threatening bleeding can occur. Factor XIII concentrates prevent bleeding. This disorder is extremely rare. For these reasons, a person with Factor XIII Deficiency must be medically followed in a treatment centre that specializes in bleeding disorders, such as a Hemophilia Treatment Centre.

For More Information

You can obtain a list of Hemophilia Treatment Centres by contacting the National Office of the Canadian Hemophilia Society:

**National Office
Canadian Hemophilia Society
625 President-Kennedy Avenue
Suite 1210
Montreal, Quebec H3A 1K2
Tel.: (514) 848-0503
Toll-free: 1-800-668-2686
E-mail: chs@hemophilia.ca
Web site: www.hemophilia.ca**

This brochure provides general information only. The CHS does NOT practice medicine and does not suggest specific treatments. In all cases, we suggest that you speak with a doctor before you begin any treatment.

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FACTOR XIII
DEFICIENCY

AN INHERITED
BLEEDING DISORDER

AN INFORMATION
BOOKLET

Canadian Hemophilia Society
We're all related  by blood.

