FACTOR VII
DEFICIENCY

AN INHERITED BLEEDING DISORDER

AN INFORMATION BOOKLET

Acknowledgements

Claudine Amesse
Nurse Coordinator, Hemophilia Centre,
Hôpital Ste-Justine,
3175, chemin Côte Ste-Catherine,
Montréal, Québec H3T 1C5

Sylvie Lacroix
Nurse Coordinator, Hemophilia Centre,
Quebec Reference Centre for the Treatment of Patients with Inhibitors
Hôpital Ste-Justine,
3175, chemin Côte Ste-Catherine,
Montréal, Québec H3T 1C5

Ginette Lupien
Nurse Coordinator, Hemophilia Centre
Hôpital St-Sacrement,
1050, Chemin Ste-Foy,
Québec City, Québec G1S 4L8

Canadian Association of Nurses in Hemophilia Care
Association Canadienne des Infirmieres et Infirmiers en Hemophilie

Members of the Canadian Association of Nurses in Hemophilia Care (CANHC)

Edited by David Page

We are very grateful to the following people, who kindly undertook to review the information in this booklet. Their suggestions were very much appreciated.

Dr. Georges-Étienne Rivard
Dr. François Jobin
Dr. Rochelle Winikoff
Gisèle Bélanger, R.N.
Claude Meilleur, R.N.
Christine Bouchard, R.N.

We would also like to thank Ms. Tiziana Fastidio for her contribution in typing and editing the French version of this booklet.

We are equally grateful for the translation from French to English by Diane Bisonnette.

Baxter

We would like thank Baxter for providing funding for the publication of this booklet.

First Edition, June 2001

PREFACE

We are pleased to present the first edition of the information booklet *Factor VII Deficiency: An Inherited Bleeding Disorder.*

This booklet has been written in order to inform people with Factor VII Deficiency and their families about the disorder, and to educate the general public.

The information presented in this document was accurate at the time of its publication. The authors and editors do not assume responsibility for any problems that may arise related to its practical clinical application.

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Introduction

Learning that you have Factor VII Deficiency is not easy. Learning your child has this disorder can even be more difficult. Feelings of insecurity and frustration are common. To make it even more difficult, Factor VII 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 VII 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.

This booklet deals with the most important questions that pertain to Factor VII Deficiency and has two main goals. The first is to describe the disease and its treatment. The second and more practical goal is to teach how to recognize a serious bleed and to take emergency measures.

This booklet will not provide you with all the answers you need. As medical care must always be personalized, your best source of information is the comprehensive care team at your Hemophilia Treatment Centre. Nevertheless, it can serve as a reference that will allow you to better understand your disease and alert you to signs or symptoms of bleeds as they occur.

How Factor VII Deficiency is Inherited

Factor VII 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 30,000 units called genes. These genes determine such things as the colour of a person's eyes. In the case of Factor VII Deficiency, one of the genes on chromosome 13 has a defect.

The defective gene in Factor VII Deficiency is on a chromosome that does not decide the sex of the child. This means that Factor VII Deficiency can affect females as well as males. In this way, it is unlike other bleeding disorders such as Factor VIII Deficiency, also called hemophilia A, in which the defective gene is sex-linked – and therefore primarily affects males.

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 VII Deficiency, both parents must be carriers. In such a case, the child inherits two defective Factor VII genes; one from the mother and the other from the father.

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

The three figures below illustrate how Factor VII Deficiency can be passed on.

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

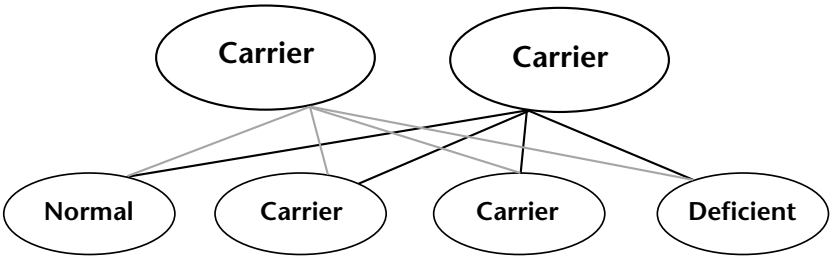

Figure 1

Figure 2 shows what can happen when someone with severe Factor VII Deficiency has children with a non-carrier. All the children will be carriers, but none of them will have the disease. (3, 13)

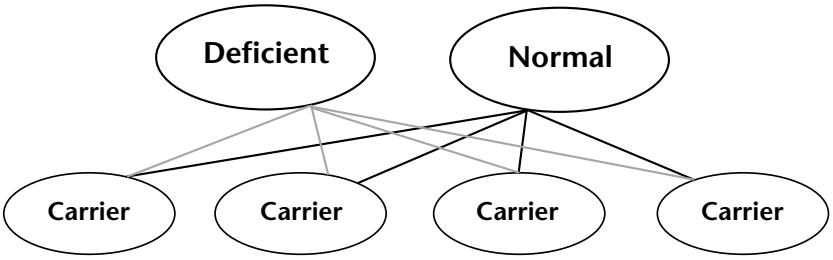

Figure 2

Figure 3 shows what can happen when someone with severe Factor

VII 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 VII Deficiency.

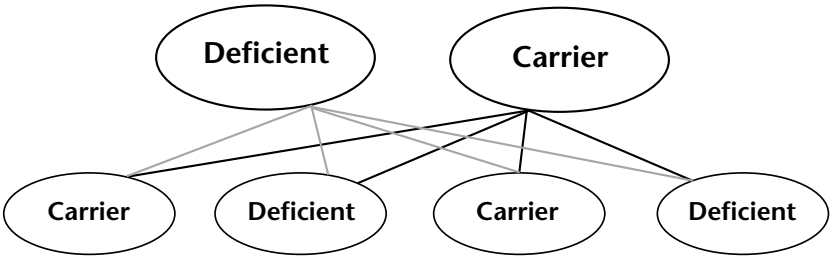

Figure 3

Incidence

Approximately 1 out of 1000 persons is a carrier of the defective Factor VII gene. However, because both parents need to be carriers of the defective gene in order to pass on the disease, severe Factor VII Deficiency is extremely rare—it occurs in 1 in 500,000 people. The disease affects males and females in equal numbers. (15)

The Degree of Severity of Factor VII Deficiency

The severity of Factor VII Deficiency depends on the level of Factor VII in the bloodstream. If Factor VII is completely absent or present at a very low level, a person has a severe deficiency. (5) The lower the Factor VII level, the greater the severity of symptoms of the disease the person has and vice versa. On the other hand, a person with a mild or moderate deficiency often has no symptoms of bleeding at all.

There are several genetic variations of Factor VII Deficiency. These variations explain why certain people have severe bleeds, mainly due to nearly absent Factor VII. (15)

However, even people with only a mild or moderate deficiency (2% to

10% of the normal level of Factor VII) can suffer from bleeds. As they do not have many bleeding problems, they may have difficulty identifying the signs and symptoms of a hemorrhage. It is equally important that these people receive care from a centre that specializes in bleeding disorders.

If you are a person with Factor VII Deficiency, even if it is mild, you must remain tuned in to your own body and immediately report any suspicious symptoms to your Hemophilia Treatment Centre. The nurse will help you to identify the exact problem and what is happening to your body.

The Cause of Bleeding in Factor VII 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 that call for help from other platelets and from clotting factors.

• The clotting factors, including Factor VII, are tiny plasma proteins. The

strands of fibrin join together to weave a mesh around the platelets. This prevents the platelets from drifting back into the blood stream. *(See Figure 4.)*

Diagnosis

Factor VII Deficiency is often

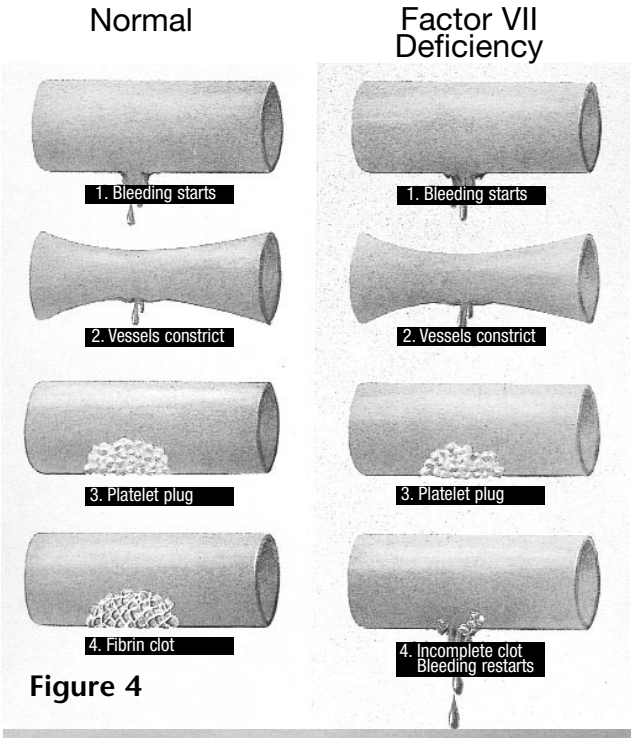

Figure 4

• Factor VII is a trace protein found in the blood. It plays a role in the coagulation cascade, the chain reaction that is set in motion to form fibrin when there is an injury to a blood vessel. Factor VII is activated, or "turned on", by tissue factor. It is turned into Factor VIIa (the "a" stands for "activated"). Factor VIIa in turn activates Factor X (pronounced Factor 10) and Factor IX (pronounced Factor 9), allowing the clotting process to continue. If one of the clotting proteins such as Factor VII is absent, the chain reaction is broken, and clotting occurs more slowly, or not at all. *(See Figure 5.)*

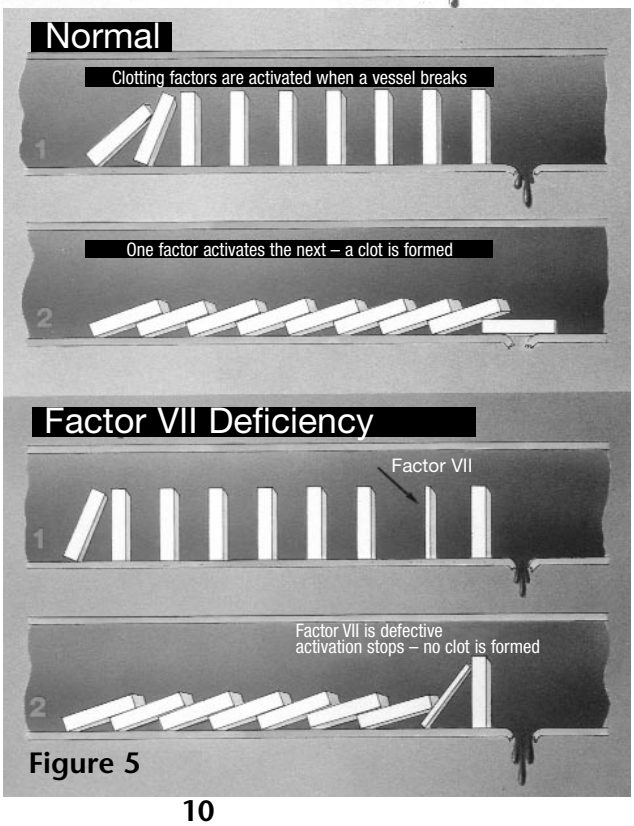

Figure 5

diagnosed when a newborn has a bleeding episode soon after birth, possibly following circumcision. (5) The diagnosis is made by measuring the level of Factor VII in the blood.

However, in some cases, Factor VII Deficiency is diagnosed later in childhood or even in adulthood. When a patient shows signs of abnormal bleeding, his/her doctor normally asks for a small blood sample in order to measure the length of time it takes for a clot to form. One of the things this test can show, especially when it is prolonged, is a deficiency in Factor VII.

Symptoms

Bleeding sites

The most common sites of bleeding with Factor VII Deficiency are:

• the joints. This is called *hemarthrosis.* Frequent bleeding into a joint can cause permanent damage to the joint, called *hemophilic arthropathy.*

• the muscles and body tissues. These bleeds are called *hematomas.* They, too, can cause serious damage, if left untreated.

• the central nervous system – the brain or the spine. This can happen after an injury or for no apparent reason.

• the mouth, usually after dental surgery or a tooth extraction. This type of bleeding is quite common in Factor VII Deficiency. Depending on the severity of the deficiency of Factor VII, affected persons may need to be treated with factor replacement therapy before any dental treatment so as to avoid bleeding. (*See Treatment on page 17.)*)

Surprisingly, bleeding during or after surgery is quite rare in people with Factor VII Deficiency, even among those whose deficiency is severe enough to cause frequent joint bleeds.

Several studies have shown that up to 16% of people with severe Factor VII Deficiency have had a bleed in the Central Nervous System, either the brain or spine. This means that good care in a treatment centre that

specializes in bleeding disorders is of vital importance. Intracranial bleeding can also occur in newborns due to trauma at birth. (1, 7, 8, 14, 16, 18)

Here is a list of the types of bleeding that may be seen in Factor VII Deficiency.

- *Menorrhagia* (excessive menstrual bleeding)
- Nose bleeds (also called *epistaxis*)
- Gum bleeding (*gingivorrhagia*)
- Gastro-intestinal bleeding (black stools, bright red vomit)
- Joint bleeds (*hemarthroses*)
- Frequent and easy bruising
- Bleeding into muscles or tissues (*hematomas*)
- Blood in the urine (*hematuria*)
- Bleeding into the brain or spine (central nervous system)
- *Retroperitoneal* bleeding (accumulation of blood in the lower posterior abdomen) (9)

Please Note:
The sections of this booklet related to…
- **Signs and Symptoms of Specific Bleeds**

- **Basic First Aid to Treat Bleeding**
- **Prevention and Healthy Living**
- **Preventing Bleeds**

have been copied from a publication of the *Canadian Hemophilia Society: The Inhibitor Guide and Notebook.* (19)

Signs and Symptoms of Specific Bleeds

A person with Factor VII Deficiency must be aware of the signs and symptoms of specific and potentially life threatening bleeds.

Bleeds that affect the head, neck, thorax (chest) or abdomen (belly) can be life threatening and require immediate medical treatment. 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 of the body organs essential to life. A bleed to the head is very serious.

Watch for:

• headache
• problems with vision (eyesight)
• nausea and vomiting
• personality changes
• sleepiness
• loss of balance***
• loss of fine motor coordination (clumsiness)***
• fainting***
• convulsions***

***These symptoms occur later in the case of a serious head injury.
If any of these symptoms occurs, consult your doctor immediately.

Neck

The tissues of the nose, 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 fill with blood, they increase pressure on the respiratory tract. This makes breathing difficult and can even block the airway completely.

Watch for:

- 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 tissues traps blood inside the air sacs that normally hold oxygen. This makes breathing difficult.

Watch for:

- 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.

Watch for:

• 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, seek immediate medical treatment.

There are other types of bleeding which require medical treatment but which are not as serious. These are described 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. (Nurses call these four treatments RICE.)

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.

Warning about muscle bleeds

Bleeding into muscles does not seem to happen very often in people with Factor VII Deficiency. Nevertheless, it is important to know the following.

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.

Treatment

Treatment with blood products or blood substitutes can temporarily raise Factor VII levels high enough to stop bleeding. This is called *factor replacement therapy*. Factor VII concentrates can be infused:

- at the time of surgery
- to the mother during and after childbirth
- for serious bleeding in a joint
- after trauma
- before dental surgery or a tooth extraction
- for any other serious bleeding episode

Currently, the most commonly used treatment in Canada is Factor VII concentrate. This factor concentrate is made from human plasma and contains only Factor VII.

Advantages

• The risk of blood clots in the bloodstream is low.

• It has been shown to be effective in controlling bleeding in Factor VII Deficiency.

• It is treated to kill known viruses such as HIV, hepatitis B and hepatitis C.

Disadvantages

• Factor VII Concentrate is made from human plasma. As a result, the risk of transmitting new, unknown infections cannot be ruled out.

The use of other products like fresh frozen plasma, Prothrombin Complex Concentrates and Recombinant Factor VIIa can sometimes be considered.

Problems Specific to Women

Menorrhagia

Women with Factor VII Deficiency may have heavy and prolonged menstrual bleeding which can be controlled using oral contraceptives. (6) If this method is ineffective, intravenous treatment with Factor VII concentrate may be used during menstruation.

Bleeding during childbirth

During a normal pregnancy, the level of Factor VII increases slightly in all women. It may be assumed that the same increase in levels occurs in pregnant Factor VII deficient females. This slight increase during pregnancy does not, however, remove the potential risk of a significant hemorrhage.
A pregnant woman with Factor VII Deficiency must be followed by a centre which is familiar with the disorder.

One medical journal article described the infusions of Factor VII concentrates every 6 hours for 72 hours during delivery to maintain the Factor VII level at 10%. This method resulted in uncomplicated deliveries with minimal post-partum bleeding. (17)

The Best Advice…

• The patient, or the parents of the patient, with severe Factor VII Deficiency (factor levels of 2% or less) must learn to recognize the signs and symptoms of severe bleeding.

• They must have immediate access to Factor VII concentrate for easy control of bleeding episodes, in the event of a life-threatening bleed (e.g. head, neck, gastrointestinal).

• Ideally, the patient, or the parents of the patient, should have enough Factor VII concentrate available for one or two treatments in case of emergency.

• If the patient is able to do so, he/she must self-infuse with Factor VII concentrate at the first sign of hemorrhage. Then, he/she must seek help at the closest hospital or Hemophilia Treatment Centre for further treatment.

• An affected person who does not self-infuse must go immediately to his treatment centre with his/her Factor VII concentrate. Once there, the personnel, who are aware of his situation, can infuse the Factor VII concentrate rapidly.

• If the patient does not keep Factor VII concentrates at home, he/she should have a letter, signed by his/her treating physician, explaining his/her condition and the emergency medical treatment required.

• A patient who has suffered from a central nervous system bleed, or who has developed a target joint (repeated bleeds into the same joint) should discuss the benefits of prophylactic treatment (regular Factor VII

concentrate treatments) with his/her doctor. The goal of prophylactic treatment is to prevent bleeds from recurring in the same sites.

Prevention and Healthy Living

Once again, we recommend the Canadian Hemophilia Society's *The Inhibitor Guide and Notebook* for more information on prevention and healthy living.

Physical Activity and Exercise – Benefits and choices

The preferred type of physical activity varies with each individual. Children usually like games. Adults prefer more structured fitness activities. People of all ages need regular exercise. It helps to prevent bleeds and to reduce the risk of complications that may occur following a bleed.

Each person's situation needs to be assessed individually. The comprehensive care team and the patient (and the parents, in the case of a child) must discuss together the benefits and the risks of various physical activities.

Choices must be made with consideration to age, the person's and family members' preferences and the person's abilities and health status. One must be prudent in choosing a physical activity; however, over-protectiveness must be avoided so as to allow the person with a bleeding disorder to undergo normal development.

Protective gear, such as a helmet, is necessary while doing certain activities. Whenever possible, sports shoes with solid support for the ankle and foot should be worn. Contact sports, such as boxing, football and hockey, should be avoided due to the risk of serious bleeds.

Activities for all ages

All ages

• The best activity is swimming. This excellent activity has few risks; it helps strengthen muscles and increases flexibility and coordination. After a bleeding episode, swimming can help to recover normal activity more rapidly.

Pre-schoolers

• Activities that do not put much stress on joints (knees, ankles and elbows) are recommended.

• Riding a tricycle, ball throwing and swimming are recommended sports.

School-aged children

• Parents must communicate with their child's teachers to educate them about Factor VII Deficiency and the role of the comprehensive care team. Teachers will find answers to their questions and their concerns, and be better able to plan safe physical activities. The school must have access to emergency resource names and phone numbers in case the child is injured.

• The child must participate in school activities and in physical education classes to ensure he/she learns to socialize normally with his/her classmates.

Teenagers and Adults

• A person with a bleeding disorder must always be aware of his/her limits and capabilities when choosing a sports activity.

• Those who wish to participate in new activities or new sports should read the chapter entitled *Staying Healthy* in the Canadian Hemophilia Society publication *All About Hemophilia*. The section on sports ranks activities according to their benefit in promoting fitness and their likelihood of causing bleeding.

Preventing Bleeds

Here are a few effective ways to prevent a bleed.

• Stay fit. Strong muscles support joints and contribute to reducing the number of joint bleeds.

• Always use appropriate gear for the sporting activity; for example, helmet, knee and elbow pads, running shoes with good ankle support and shock absorbent heels.

• Rest after a joint or muscle bleed to help healing and prevent another bleeding incident.

• Avoid alcohol. Alcohol affects platelet function. This can cause bleeding to occur more easily.

• Prevent dental problems and gingivitis. See the dentist every six months. Your centre can recommend a dentist who is familiar with hemophilia.

• Never take aspirin (ASA); use alternate medications recommended by your centre.

• Check with your Hemophilia Treatment Centre before taking any new drug. This includes all organic and vitamin supplements without a DIN number.

Keep in mind

• Check your joints daily.

• Eat healthy foods in adequate amounts to stay fit. Being overweight increases the stress on knees and ankles and may augment the risk of a bleed in these joints.

• Notify your treatment centre team prior to having dental extractions or dental surgery. They will ensure that you receive adequate factor replacement therapy, if needed.

• Wear a MedicAlert bracelet that describes your bleeding disorder at all times. Remember that Factor VII Deficiency is very rare. Medical personnel who are not familiar with you will need the support of the

information provided by MedicAlert.

• Inform your comprehensive care centre of any trip you plan. Obtain a travel letter from your doctor. Obtain an adequate amount of factor concentrate and bring this manual for reference.

• Keep a photocopy of your medical insurance card in your manual.

• Always stay in contact with your Hemophilia Treatment Centre.

Vaccination

The Canadian Pediatric Association has published several documents promoting the Canadian immunization program. At an early age, the child should receive the basic vaccinations. However, certain precautions should be taken to avoid a bleed at the site of the injection. The nurse at the Hemophilia Treatment Centre can inform you of the precaution you must take .

Special considerations

A child with Factor VII Deficiency should be vaccinated on a timetable set by his/her pediatrician or family doctor. However, anyone receiving blood products of human origin should be vaccinated against hepatitis A and hepatitis B.

Hepatitis A has been transmitted, on very rare occasions, 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) are at 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.

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 have symptoms 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 Havrix. 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, usually a hematologist
- 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.

Conclusion

Factor VII Deficiency is a rare hereditary bleeding disorder. It affects both men and women. In people who suffer from a severe deficiency, it can cause serious bleeding. There are Factor VII concentrates available which allow bleeding episodes to be controlled. It is important that your health care be provided by a centre that is familiar with the care of people with bleeding disorders.

For More Information

You can obtain a list of Hemophilia Treatment Centres by contacting the

National Office of the Canadian Hemophilia Society at:

Canadian Hemophilia Society
625 President Kennedy Ave., Suite 1210
Montreal, Quebec
H3A 1K2
(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 medecine 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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