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abnormal gene When a gene has a mistake in its structure, it is called a mutant gene or abnormal gene.

acetaminophen A drug used to relieve pain. A common brand name is Tylenol®. Acetaminophen does not affect platelet function and is safe for use in people with hemophilia.

acetylsalicylic acid (ASA) The active ingredient in Aspirin® and many other over-the-counter pain relievers and cold remedies, such as Entrophen®, Anacin®, Norgesic®, 222's, Dristan®, 282's, 292's, Coricidin Cold®, Coricidin D®, Robaxisal® and Midol®. This drug affects platelet function and should not be used in people with hemophilia.

activated partial thromboplastin time (aPTT) A basic screening test for clotting problems. However, it is not very reliable in the diagnosis of mild and moderate hemophilia A.

activated prothrombin complex concentrates Plasma-derived concentrates that contain many activated clotting factors. These activated clotting factors can "bypass" an inhibitor's actions. FEIBA® VH is a brand name often used in Canada.

acute bleed A bleed which is currently in progress.

acute pain In hemophilia, pain which is caused by acute bleeding and not by a chronic joint disease such as arthritis.

acute synovitis A condition in which the lining of the joint (synovium) remains inflamed for several weeks after a joint bleeds.

AHCDC The Association of Hemophilia Clinic Directors of Canada.

AIDS Acquired Immune Deficiency Syndrome, caused by the human immunodeficiency virus (HIV). This virus was transmitted in certain blood products before 1987.

albumin A protein found in human plasma and used to treat shock and burn victims. It may be used to stabilize factor VIII in certain recombinant factor concentrates. The currently available recombinant factor VIII concentrates use sucrose, rather than albumin, as a stabilizer.

allergic reactions Very infrequently, clotting factor concentrates may cause allergic reactions. Allergic reactions can be mild, such as hives or rash, or severe, such as wheezing or *anaphylaxis* (difficulty breathing). Severe reactions need emergency treatment.

amniocentesis A test used to detect abnormalities in the fetus. A thin needle is inserted through the abdomen and into the uterus to obtain a small amount of amniotic fluid. The amniotic fluid contains cells shed by the fetus, which are examined for defects.

anaphylaxis A sensitivity reaction triggered by food or medicine characterized by difficulty breathing, nausea and vomiting among other symptoms.

anemia A condition caused by low hemoglobin levels because of blood loss, leading to fatigue and lack of energy.

antibody A natural chemical substance produced in the blood by the body's immune system to defend against harmful substances. Antibodies that occur in people with hemophilia are called inhibitors.

anti-diuretic A substance that makes the body retain water. Desmopressin acetate, used in the care of mild hemophilia A, has this side effect.

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antifibrinolytics Drugs (Cyklokapron®) that help to hold a clot in place once it has formed by stopping the activity of an enzyme, called plasmin, which dissolves blood clots. Antifibrinolytic drugs prevent the rapid breakdown of blood clots, a natural bodily process that appears to be increased in women with heavy menstrual bleeding.

anti-hemophilic globulin The first blood product preparation containing high concentrations of factor VIII, discovered in the 1930s.

arthritis Inflammation of the joint. In addition to inflammation of the synovial lining, there is also damage to the cartilage and bones of the joint surfaces. In hemophilia, arthritis is caused by repeated bleeding into the joint cavity.

arthroscopic synovectomy An operation to remove the inflamed synovium in a joint. A tube (arthroscope) is inserted into the joint through a small incision. Special tools allow the surgeon to remove the thickened synovium. This operation can also be called a closed synovectomy.

Bethesda assay A test to measure the level of a clotting inhibitor once it is known to be present. The results of the test are given in Bethesda Units (BU).

Biologics and Genetic Therapies Directorate The Health Canada agency that is responsible for the licensing and safety of blood and blood products, including recombinant factor concentrates.

bleed diary A record of each bleed, including the site of the bleed, the type of clotting factor concentrate infused, the quantity infused, the lot number and the date. The diary is completed by the caregiver or the person with hemophilia himself, and returned to the HTC on a regular basis. It is also called a treatment diary or home infusion diary.

bleeding disorder A disease in which the body is unable to form blood clots as quickly or as effectively as normal. The family of bleeding disorders includes hemophilia A, hemophilia B, von Willebrand disease, platelet function disorders and a variety of rare factor deficiencies. The disorder may be hereditary or acquired.

blood clotting The process of forming a permanent clot to repair a damaged blood vessel. It includes four steps: vasoconstriction, platelet aggregation, platelet adhesion, and the formation of a fibrin plug.

blood clotting proteins Substances that circulate in the bloodstream, necessary for blood clotting. They include von Willebrand factor, and factors I, II, III, V, VII, VIII, IX, X, XI and XIII.

blood donor screening and testing A series of measures used to ensure that blood donations are safe. These include strict guidelines for donor selection, a donor questionnaire and a series of tests to look for signs of infection by HIV, hepatitis B, hepatitis C and other pathogens.

blood platelets Tiny cell fragments, less than 1/10,000 of a centimetre in diameter, circulating in the blood. There are 150 to 400 billion platelets in a normal litre of blood. Platelets are first to arrive at the site of a break in a blood vessel wall. They play an important role in stopping bleeding by clumping together, thereby beginning the repair of injured blood vessels.

breakthrough bleeding Bleeding that occurs while on treatment such as prophylaxis.

bypassing therapy A treatment for patients with inhibitors. The factor concentrate infused contains clotting factors that work around the inhibitor. In Canada, the most commonly used bypassing therapy is called FEIBA® VH.

CANHC Canadian Association of Nurses in Hemophilia Care.

carrier A female who has an abnormal X chromosome carrying the hemophilia gene. One of her two X chromosomes has a mutation of the factor VIII or factor IX gene, resulting in decreased levels of clotting factor VIII or IX. See *obligate carrier, possible/potential carrier, and symptomatic carrier*.

carrier testing A series of tests used to find out if a woman is a carrier of hemophilia. They can include both coagulation testing and DNA testing.

cartilage A flexible connective tissue. In a joint, cartilage forms a protective cap at the end of the bones, provides padding that allows the joint to move smoothly and without pain.

Centre for Infectious Disease Prevention and Control The Health Canada agency that works to ensure the safety of blood products by surveillance work on old and new diseases that could infect blood and blood products.

central venous access device A surgical implant that allows easier access to a vein for infusion of factor concentrates. This device is sometimes called a port-a-cath.

chorionic villus sampling A type of pre-natal testing for hemophilia. A very small sample of the chorionic villus (part of the placenta) from inside the womb is taken out and tested in the lab.

Christmas Disease Another term for hemophilia B or factor IX deficiency, named after Stephen Christmas, a Canadian with hemophilia who was the first person to be diagnosed with the disorder.

chromosome A long chain of chemicals known as DNA, which is arranged into hundreds of units called genes. Genes determine such things as the colour of a person's eyes.

chronic pain In hemophilia, pain due to damaged tissues and/or altered brain and spinal cord functions and not an acute bleed.

CHS Canadian Hemophilia Society.

CJD Creutzfeldt Jakob Disease, a fatal brain disease believed to be caused by infection with a misshapen protein, called a prion. There is no evidence that the classical form of CJD is transmitted by blood or blood products.

classical hemophilia Another term for hemophilia A or factor VIII deficiency.

closed synovectomy See *arthroscopic synovectomy*.

clotting factor concentrate A lyophilized preparation of clotting proteins, which is dissolved in sterile water for infusion to correct a coagulation disorder. The concentrates can be manufactured from human plasma or by recombinant technology. Concentrates exist to correct deficiencies in factors I, II, VII, VIII, IX, X, XI, XIII and von Willebrand factor.

clotting factor modification A process by which the factor VIII or IX molecules are intentionally modified to make them better for the treatment of hemophilia, for example, by extending their half-life.

clotting factor purification The manufacturing steps whereby factor VIII or IX proteins are separated from other substances including bacteria and viruses in a series of physical and chemical purification processes.

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clotting factor recovery The amount of clotting factor concentrate a person's body can actually use to stop bleeding compared to the amount infused.

clotting factor therapy The treatment in which clotting factors are infused into the bloodstream of a person with hemophilia to replace those that are missing, and temporarily correct the coagulation disorder. Also called *factor replacement therapy*.

coagulation A complex process that makes it possible to stop torn blood vessels from bleeding. The four stages in the coagulation process are vasoconstriction, platelet adhesion, platelet aggregation and the formation of a fibrin plug by clotting factor proteins.

coagulation cascade The chain reaction in which clotting factors, which are tiny plasma proteins, link to form a chain, called fibrin, around the platelets at the site of a break in a blood vessel wall.

coagulation laboratory A laboratory which is specialized in doing the many tests needed to correctly diagnose the different coagulation disorders, including hemophilia A and B.

coagulation testing The many tests needed to correctly diagnose the different coagulation disorders, including hemophilia A and B.

compartment bleed A deep bleed inside a closed-in space, such as the forearm, calf or iliopsoas muscles. Compartment bleeds are serious because they can cause damage to important nerves and blood vessels.

comprehensive care All of the medical services needed by a person with hemophilia and his/her family for the treatment of hemophilia and related conditions. This care is provided at a hemophilia treatment centre.

comprehensive care team The team of people involved in the care of a person with hemophilia. They include a medical director, nurse coordinator, physiotherapist, social worker and parent/caregiver. Other health professionals are added to the team as needed.

corpus luteum cyst A type of cyst that can form on an ovary if bleeding occurs during ovulation, when the egg is released from its follicle. See *hemorrhagic ovarian cyst*.

cryoprecipitate A blood component made from plasma, containing factor VIII, commonly used to treat hemophilia A in the past. However, because there is no method to kill viruses in cryoprecipitate, it is no longer recommended to treat factor VIII deficiency.

Cyklokapron® An antifibrinolytic drug (tranexamic acid) that helps to hold a clot in place once it has formed by stopping the activity of an enzyme, called plasmin, which dissolves blood clots.

desmopressin acetate A synthetic drug that is a copy of a natural hormone. It acts by releasing von Willebrand factor (VWF) stored in the lining of the blood vessels. The additional VWF helps raise factor VIII levels by increasing von Willebrand factor. Desmopressin is not made from blood. It can be called DDAVP® Injection, Octostim®, Octostim Spray®, Stimate® and Stimate® Spray.

direct mutation testing A test to identify the presence of the actual hemophilia mutation.

DNA Deoxyribonucleic acid. DNA works as the building blocks of our genetic make-up. The DNA in each chromosome is arranged in thousands of units called genes. Each one of the genes directs the body to produce certain proteins, including clotting proteins.

DNA linkage analysis A type of test to determine carrier status, done if specific mutation is not known. It involves following DNA markers that are either within and/or surround the hemophilia gene. DNA linkage analysis does not identify the specific mutation or factor activity level, but it can provide information about the specific “pattern” of the factor gene mutation. This genetic pattern provides information about carrier status with a certain degree of probability.

DNA polymorphism testing A test for genetic markers (called polymorphisms) that are within or close to the gene mutations that cause hemophilia. It is the most common kind of DNA testing used today.

DNA testing A process to determine whether a female is a carrier. There are two kinds of DNA testing: DNA polymorphism testing and direct mutation testing.

dysmenorrhea Although some pain during menstruation (menstrual periods) is a common complaint among women in general, about 50 per cent of carriers experience moderate to severe menstrual pain during menstruation called dysmenorrhea.

EMLA® (Eutectic Mixture of Local Anesthetics) A cream to help decrease the pain of the needle going into a port-a-cath or vein.

endometrial ablation A surgical procedure to remove or destroy the lining of a woman's uterus. This procedure is sometimes considered for women who have abnormalities of the uterus or for carriers who have not responded to other types of treatment and who have completed childbearing and do not wish to preserve fertility.

endorphins A group of chemicals produced naturally by the body and brain that contribute to a sense of well-being. Continuous exercise of 20-30 minutes duration or more is believed to temporarily stimulate the production of endorphins.

epidural Pain control for child delivery given by regional anesthesia. Prior to giving birth, all carriers should meet with an anesthetist to discuss pain control options for the delivery.

epistaxis Bleeding from the nose.

exercise Activity that involves physical exertion, voluntary movements and burning calories, and is specifically planned, structured and repetitive. Examples include jogging, cross-country skiing, recreational swimming, cycling and aerobics.

factor IX A protein in the blood that is essential for clotting. Factor IX levels are low in people with hemophilia B.

factor IX deficiency hemophilia See *hemophilia B*.

factor VIII A protein in the blood that is essential for clotting. Factor VIII levels are low in people with hemophilia A.

factor VIII deficiency hemophilia See *hemophilia A*.

factor assay A test done to measure the level of clotting factors in the bloodstream of a person. The standard used is 100 percent. Normal people vary between 50 and 150 percent. People with severe hemophilia A or B have less than 1 percent of the normal quantity of factor VIII or IX.

factor concentrates See *clotting factor concentrates*.

factor purification See *clotting factor purification*.

factor replacement therapy See *clotting factor therapy*.

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femoral nerve A nerve running along the iliopsoas muscle that can be easily compressed when the muscle fibres swell with blood.

fibrin The material making up the clot that forms in the last stage of the coagulation process.

fibrin glue A treatment that is sprayed directly onto a surface wound to form a clot and stop bleeding. It is made of two components—fibrinogen and thrombin. It also contains an enzyme to strengthen the clot and an antifibrinolytic to prevent the clot from breaking down.

fifth disease A common childhood disease caused by infection with parvovirus B19.

frenulum The piece of skin attached to the upper lip and gum.

fresh frozen plasma A component of whole blood that was used for treatment of hemophilia in the 1950s and 1960s.

gene Tiny structures of DNA that determine such things as the colour of a person's eyes. Hemophilia is caused by an abnormal gene on the sex chromosome.

gene therapy A treatment involving the delivery of a normal copy of the clotting factor gene to the patient's cells.

genetic counselor A person who understands genetic testing and is trained to work with couples who are planning to have a baby, or who are already pregnant.

genetic disorder A disease that is caused by a gene that does not work normally. Genetic disorders like hemophilia can be passed from generation to generation.

geneticist A person who studies genes and how people inherit diseases.

genetic mutation The specific mistake in the gene.

genetics The study of how genes are passed from one generation to the next.

genetic testing Genetic testing can identify the specific factor VIII or IX gene mutation in an individual with hemophilia or a carrier, in 90 to 99 percent of cases. This type of test can be used for prenatal diagnosis to determine whether or not a fetus carries the genetic mutation.

gynecologist A physician who specializes in the female reproductive system.

half-life The time taken for half the infused clotting factor activity to disappear from a person's bloodstream.

hemarthrosis A bleed into a joint.

hematologist A physician specializing in diseases of the blood.

hematology The medical specialty dealing with diseases of the blood.

hematology laboratory A laboratory that does a wide range of blood tests.

hematoma A bleed into tissues or a muscle, also called a bruise.

hematuria Blood in the urine, caused by bleeding in the kidneys.

hemoglobin A substance in the red cells of blood, responsible for carrying oxygen.

hemoperitoneum Bleeding into the pelvic tissues and ligaments and sometimes into the abdominal and pelvic cavity. It is a serious and possibly life-threatening situation and requires urgent medical attention.

hemophilia A term used to describe bleeding disorders caused by low levels of factor VIII or IX (hemophilia A or B).

hemophilia A A genetic disorder characterized by frequent bleeding into joints, muscles and tissues. The prolonged bleeding is caused by low levels of factor VIII. The disease is also called *classical hemophilia* and *factor VIII deficiency*.

hemophilia B A genetic disorder characterized by frequent bleeding into joints, muscles and tissues. The prolonged bleeding is caused by low levels of factor IX. The disease is also called *Christmas Disease* and *factor IX deficiency*.

hemophilia treatment centre (HTC) A medical clinic that provides comprehensive care for people with hemophilia.

hemorrhage The escape of blood from blood vessels, either on the surface of the body or internally.

hemorrhagic ovarian cyst A type of cyst (called a corpus luteum cyst) that can form on an ovary if bleeding occurs during ovulation, when the egg is released from its follicle. Carriers of hemophilia are more likely to have significant bleeding at ovulation.

hepatitis A An acute viral disease which causes liver inflammation and jaundice, usually lasting several weeks, that does not develop into chronic liver disease. It is transmitted by dirty water or contaminated food because of poor hygiene. As a precaution, it is generally recommended that all individuals with hemophilia be vaccinated against hepatitis A.

hepatitis B A viral disease which can result in chronic liver disease. It can be transmitted through sexual contact and contaminated needles. The hepatitis B vaccine is provided free of charge as part of routine childhood immunizations for all children in Canada.

hepatitis C A viral disease which in 80 percent of cases becomes chronic. It causes liver damage, which usually takes many years to develop. It is transmitted by the exchange of contaminated needles. In very rare cases it can still be transmitted by fresh blood components.

hepatologist/gastroenterologist A physician specializing in diseases of the liver.

high responder A term used to describe a person with a clotting inhibitor whose immune system reacts very strongly to infusions of factor concentrate. He develops a high titer inhibitor soon after he receives an infusion.

high titer inhibitor An inhibitor that is measured at more than 5 Bethesda Units. The antibodies of a person with a high titer inhibitor are stronger and destroy the factor concentrate more quickly.

HIV Human immunodeficiency virus, responsible for AIDS.

home care The care of the person with hemophilia at home, rather than in hospital. This includes the administration of clotting factor concentrates by the person with hemophilia or by a family member.

home infusion The administration of clotting factor concentrates by the person with hemophilia or by a family member in the home setting.

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hormonal treatment Hormones are used to treat heavy menstrual bleeding (menorrhagia) or abnormal menstrual bleeding (metrorrhagia). Hormones help decrease the thickness of the uterus lining thus decreasing menstrual bleeding. The most common hormone therapies are oral contraceptives (estrogen and progestin) or the levonorgestrel intrauterine system.

hydrotherapy Physiotherapy which uses water as resistance in the rehabilitation of muscles and joints.

hyponatremia A condition in which the sodium or salt content in the blood is low. In rare cases, this may result in a seizure occurring. It can be caused by excessive water retention, which can be a side effect of desmopressin.

hysterectomy Removal of a woman's uterus. This procedure is sometimes considered for women who have abnormalities of the uterus or for carriers who have not responded to other types of treatment and who have completed childbearing and do not wish to preserve fertility.

iliopsoas A large muscle in the pelvic region near the hip joint. Bleeding here can damage the large nerve that controls the muscles at the front of the thigh, as well as the major artery at the front of the leg.

immune tolerance induction therapy The infusion of high doses of the missing clotting factor concentrate 3-7 times per week for very long periods of time—months or years. The objective of the therapy is to allow the body's defenses to become accustomed to the foreign factor and to stop making antibodies against it, so that normal doses will be effective in stopping bleeding.

immunoabsorption A technique for people with inhibitors by which the patient's plasma is taken out of his body by intravenous access and passes through a sophisticated machine with columns. These columns remove only the antibodies from the plasma. The patient's plasma is returned to him without the antibodies, allowing clotting factor concentrates to be infused.

inflammation Swelling and redness of a part of the body, often warm to the touch and painful, such as when there is bleeding in a joint or muscle.

infusion The administration of clotting factor concentrates intravenously using a syringe and butterfly needle, or using a central venous access device, such as a port-a-cath.

inherited condition A disorder which is caused by a genetic mutation and which is transmitted from one, or both, of the parents to the child at the time of conception.

inhibitors Antibodies produced to eliminate factor VIII or IX or other clotting factor proteins, seen as foreign by the body's immune system.

international unit (IU) A standardized measurement of the amount of factor VIII or IX contained in a vial.

intravenous The infusion of a medication directly into a vein.

inversion The most common type of mutation in hemophilia is an inversion in factor VIII (half of males with severe hemophilia A have an inversion in their factor VIII gene). See also *mutation analysis*.

joint A joint is the place where two bones come together, allowing movement such as bending, rotating and swinging back and forth.

joint bleed Caused by a tear in the lining of the joint (synovium), blood escapes from the blood vessels and gradually fills the joint cavity. A joint bleed is usually because of “pinch or twist” types of injury to the joint. It can happen especially if the activity involves putting full weight and stress on a joint or forceful movements such as throwing and kicking.

joint disease Synovitis and arthritis. These diseases in hemophilia are caused by repeated bleeding into joints. They are most common in knees, ankles and elbows.

joint replacement The complete replacement of the joint cavity with synthetic materials. The joints most commonly replaced are the knee and the hip. However, the operations cannot be performed on young people as the materials used to replace the joint wear out.

joint capsule The joint space and the tissue membrane that forms a “sleeve” around the joint together form the joint capsule.

ligament Connective tissue connects two bones or cartilage or holds together bones and muscles.

long-term prophylaxis The most common type of prophylaxis presently used. Infusions of factor concentrates are given 2 or 3 times per week throughout a boy's childhood. The goal of this therapy is to prevent bleeding and avoid joint damage.

low responder A term used to describe a person with a clotting inhibitor whose level does not rise above 5 Bethesda Units even if he receives clotting factor therapy. Regular factor concentrates may be used to control bleeding. However, he might have to be infused more often and with higher doses.

low titer inhibitor An inhibitor that is measured at less than 5 Bethesda Units.

lyonization A genetic process whereby either one of a carrier's two X chromosomes is randomly inactivated during development. It is also called X-inactivation.

mandibular block Freezing of the lower jaw.

medical director A key member of the comprehensive care team. The medical director of a hemophilia treatment centre is usually a hematologist. He/she oversees the comprehensive care team, suggests treatments to control and prevent bleeding and oversees patients' health.

menorrhagia Heavy bleeding during a menstrual period and/or prolonged menstrual bleeding. It is one of the most common gynecological symptoms experienced by carriers of hemophilia. Menstrual bleeding can be heavy in terms of the amount of blood loss during the period and/or due to a prolonged menstrual period (more than seven days).

metrorrhagia Abnormal/irregular bleeding that occurs beyond the normal menstrual period. The abnormal bleeding occurs sometime between the end of one menstrual period and the beginning of the next, and the duration of bleeding and amount of blood loss can vary.

mid-cycle abdominal pain (mittelschmerz) Abdominal pain that occurs midway in the menstrual cycle during ovulation, when the ovaries release a new egg into the fallopian tubes. Mid-cycle abdominal pain is called mittelschmerz (German for “middle pain”). Carriers of hemophilia are more likely to have mittelschmerz than non-carriers due to bleeding at ovulation.

mild hemophilia A genetic coagulation disorder characterized by bleeding after trauma or surgery. The level of factor VIII or IX in the bloodstream is from 5 to 30 percent of normal.

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moderate hemophilia A genetic coagulation disorder characterized by bleeding after minor injury, more serious trauma or surgery. The level of factor VIII or IX in the bloodstream is from 1 to 5 percent of normal.

mucous membrane An extension of the skin inside the body—for example, the insides of the mouth, the nose, the intestines (the gut) and the uterus (the womb).

muscle A muscle is a band or bundle of tissue and fibers that have the ability to contract — this produces movement such as lifting, standing up and sitting down, walking, etc.

muscle bleed Bleeding into a muscle, which can be caused by a single serious trauma or small but repetitive trauma. A muscle bleed can happen if a muscle is strained or stretched too much.

musculoskeletal Relating to or involving the muscles and the skeleton.

mutant gene A gene with a mistake in its structure.

mutation The specific mistake in the gene.

mutation analysis A test involving laboratory analysis of the genes responsible for hemophilia. The test looks for changes in either the factor VIII or factor IX gene. If the specific gene mutation in the person with hemophilia is known, accurate carrier testing by mutation analysis can be carried out on female family members.

NHF The National Hemophilia Foundation in the United States.

nurse coordinator A key member of the comprehensive care team. Usually she/he is the coordinator of the comprehensive care team. She/he schedules appointments, answers patients' telephone calls, performs infusions at the clinic and teaches people about hemophilia.

obligate carrier All daughters of men with hemophilia will inherit the hemophilia factor VIII or IX genetic mutation. They are called obligate carriers.

on-demand therapy An infusion of clotting factor concentrate as soon as the person with hemophilia, or a parent, is aware of a bleed. The goal is to promptly stop the bleed.

open synovectomy See *surgical synovectomy*.

orthopedic surgeon A doctor who treats bones and joints.

parvovirus B19 A human virus carried by a large percentage of the population, responsible for fifth disease. Normally harmless, it can, in rare cases, cause leukemia and anemia. It can also cause miscarriage.

pediatrician A doctor who treats children from infancy to age 18. She/he has a lot of experience with diseases and health problems that affect children.

physical activity Any activity that involves physical exertion and voluntary movements that burn calories. For examples include gardening, dancing, walking the dog, shoveling snow and raking leaves.

physiotherapist A key member of the comprehensive care team. The physiotherapist is a person who is trained to keep a person's muscles and joints healthy. She/he can give advice on how to prevent or limit bleeding. She/he can help patients to understand what a bleed is, whether a bleed is serious or not and what to do to get better after each muscle or joint bleed. The physiotherapist can also give advice on how to be active and physically fit.

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physiotherapy The use of exercise to stay fit or rehabilitate weakened muscles and damaged joints.

plasma The component of whole blood that contains clotting factor proteins, including factor VIII and IX, as well as immunoglobulins and albumin.

plasma-derived clotting factor concentrate A lyophilized preparation of factor proteins, manufactured from human plasma, which is dissolved in sterile water for infusion to correct a coagulation disorder.

plasmapheresis A technique for people with clotting inhibitors by which the patient's plasma, which contains the inhibitor, is removed using specialized equipment, and replaced with albumin and/or plasma. While the inhibitor level is low after his plasma has been replaced, factor concentrates can be given for a short period of time.

platelets Small cells less than 1/10,000 of a centimetre in diameter circulating in the blood, which stick to and spread on the walls of the damaged blood vessel to promote clotting.

platelet adhesion The clumping together of platelets at the site of a tear in a blood vessel wall.

platelet aggregation By emitting chemical signals calling for help from other platelets and from clotting factors, like factor VIII and factor IX, the spreading platelets release substances that activate other nearby platelets which then clump at the site of blood vessel injury to form a platelet plug.

point mutation A type of gene mutation that causes a severe form of hemophilia.

polymorphism A genetic marker that is within or close to the gene mutation that causes hemophilia.

port-a-cath A central venous access device that is surgically implanted just under the skin. It allows easier infusion of clotting factor concentrates if access to the veins is more difficult, as it sometimes is with small children.

post-partum bleeding Bleeding after childbirth. In general, the risk of hemorrhage within the first 24 hours following childbirth is four to five percent for non-carriers. This risk increases to 22 percent for carriers.

possible carrier All daughters of carriers have a 50% chance of inheriting the hemophilia mutation. They are called possible carriers.

pre-natal testing Testing carried out to determine if the fetus is affected by hemophilia or not. Two kinds of tests can be done: chorionic villus sampling (CVS) and amniocentesis.

prophylaxis Regular infusions of clotting factor concentrates, usually 2 to 3 times a week. This is done in order to prevent bleeding episodes from happening.

psychologist A person who is a health professional who specializes in the human mind and how it functions.

quarantine The holding back of a blood product or other drug for a short period of time because of a possible problem with its quality. During this time, manufacturers and Health Canada investigate. If the problem is found to be real, the product is recalled or withdrawn. If it is found that there is no problem, the product is released from quarantine.

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radioactive synovectomy An operation to shrink the synovium in a joint. A radioactive chemical is injected into the joint. The swollen synovium dies and is eventually replaced by a healthy one.

recall The removal of a blood product or other drug from the market because the manufacturer or the regulator, Health Canada, has reason to believe the product is not safe for use.

recombinant clotting factor concentrate A lyophilized preparation of factor proteins, manufactured by recombinant technology, which is dissolved in sterile water for infusion to correct a coagulation disorder.

recovery The amount of clotting factor concentrate a person's body can actually use to stop bleeding compared to the amount infused.

rheumatologist A doctor who treats health problems and diseases that cause pain and swelling in and around muscles and joints.

self-infusion The administration by the person with hemophilia himself of clotting factor concentrates. This is done intravenously using a syringe and butterfly needle.

severe hemophilia A genetic coagulation disorder characterized by frequent bleeding episodes and bleeding after minor injury, more serious trauma or surgery. The level of factor VIII or IX in the bloodstream is less than 1 percent of normal.

short-term prophylaxis Regular infusions of clotting factor concentrates given for several days or a few weeks. Short-term prophylaxis can involve factor infusions given to control bleeding during and after surgery, or for a joint that has had frequent bleeding within a short period.

social worker A key member of the comprehensive care team. The social worker's job is to help parents, siblings and the child himself deal with the psychological, social and potentially even financial impacts that hemophilia can have on their lives.

sporadic hemophilia An isolated case of hemophilia that appears to occur in that family for the first time. Many of these new cases of hemophilia are due to new changes in the human genetic code.

sports Physical activity that involves a set of rules, or goals to train and excel in specific athletic skills. Some are individual sports such as golf and swimming. Others are played in teams—for example, soccer and hockey. Sports are often, but not always, competitive.

steroid injections Medication used to speed healing in a joint. After an infusion of factor concentrates, a small needle is used to inject the steroid directly into the joint to settle the inflammation.

superficial muscle An outer muscle as opposed to a deep or inner muscle.

surgical synovectomy An operation to remove the synovium in a joint. It is used when more extensive joint damage with cartilage erosion makes an arthroscopic synovectomy impractical. The surgeon makes an incision in the joint to remove the synovium and smooth any rough spots on the bones themselves. This operation is also called an open synovectomy.

survival study A series of blood tests over 24 to 48 hours is done to find out the half-life of factor VIII or IX in an individual.

symptomatic carrier of hemophilia A female whose factor VIII or IX level is lower than normal and who has symptoms of factor deficiency similar to mild hemophilia.

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synovectomy An operation to remove the inflamed synovial membrane in a joint. Three types of synovectomy are done: surgical (open), arthroscopic (closed) and radioactive (chemical).

synovial membrane Also called the synovium. It is a layer of special cells that line the capsule of the joint.

synovitis A type of joint disease caused by repeated bleeding into the joint cavity. The synovial membrane, in an attempt to remove old blood and debris from the joint, grows new blood vessels, becomes thicker and prone to repeated hemorrhaging.

synovium See *synovial membrane*.

target bleeding Bleeding that occurs repeatedly, over a short period of time, in the same part of the body.

target joint A joint that has had frequent bleeding in a short period of time and show signs of inflammation and loss of range of motion. A target joint is one that does not return to a healthy state between bleeds. It has a higher risk of developing early joint disease.

TENS Transcutaneous electrical nerve stimulation. This technique is used to control pain. It works by sending a very mild current of electricity to the nerves that normally send pain messages to the brain, blocking the pain signals.

termination mutation A type of genetic mutation that causes a severe form of hemophilia.

testing of blood donations A series of tests to look for signs of infection by viruses such as HIV, hepatitis B, hepatitis C, as well as other blood-borne pathogens.

thrombin A substance that aids in blood clotting, formed by a chain reaction of clotting factor proteins.

topical thrombin A powder that can be put directly on the skin or in the mouth to stop bleeding, or placed on a moist gauze or gelatin sponge and then applied with pressure. A brand name is Thrombostat®.

tranexamic acid An antifibrinolytic drug (Cyklokapron®) that helps to hold a clot in place once it has formed by stopping the activity of an enzyme, called plasmin, which dissolves blood clots.

transition A process of change and evolution from one stage to another. In hemophilia, transition refers to the continual process of learning about hemophilia and how to manage the many aspects of care. It is an ongoing team effort involving the parents, child, and comprehensive care team. See *transition to adulthood / adult independence*, *transition to adult care*.

transition to adulthood / adult independence The overall goal and journey shared by the comprehensive care team and the family, to help the child with hemophilia develop the knowledge and range of skills that he will need to manage on his own as an independent adult.

transition to adult care The coordinated transfer of a patient from child health services to adult health services, an important aspect of comprehensive care for hemophilia.

Glossary

variant CJD Variant Creutzfeldt Jakob Disease (vCJD) is the human form of Bovine Spongiform Encephalopathy, or BSE, a fatal brain disease believed to be caused by infection with a misshapen protein, called a prion. This infection is thought to be the result of eating contaminated beef products.

vasoconstriction The first stage in blood clotting in which the blood vessel constricts to reduce the flow of blood to the damaged area.

vascular constriction See *vasoconstriction*.

viral inactivation The process used to kill or eliminate viruses that might be present in plasma-derived blood products. Chemical techniques such as treatment with solvent detergents, and physical techniques such as heat treatment, filtration and other purification technologies, are used.

von Willebrand disease (VWD) A family of inherited diseases in which the blood clots more slowly than normal. The prolonged bleeding is caused by a deficiency in the von Willebrand factor.

von Willebrand factor (VWF) The clotting protein that is deficient in VWD. The VWF is either present at lower than normal levels or it does not work properly. VWF also plays an important role in transporting factor VIII in the bloodstream.

withdrawal The removal of a blood product or other drug from the market because the manufacturer, while having no reason to believe it is not safe, feels the product does not meet its manufacturing standards.

WFH World Federation of Hemophilia.

X-inactivation A genetic process whereby either one of a carrier's two X chromosomes is randomly inactivated during development. It is also called lyonization.