STEPS for LIVING
Education for all life stages.

Basics of Bleeding Disorders

What is a Bleeding Disorder

A bleeding disorder is a condition in which a person tends to bleed longer (not faster) before a blood clot forms to stop the bleeding. A bleeding disorder can be caused by a defect in the blood vessels or from an abnormality in the blood itself. The abnormality may be in blood clotting factors (proteins called coagulation factors) or in platelets (small protein-containing blood cell elements, called fragments).

What is Hemophilia?

Hemophilia is an inherited (genetically passed to the child through the parent's DNA) bleeding disorder in which the blood does not clot properly. The overwhelming majority of people with hemophilia are men and boys because of how the disorder is passed genetically from parent to child.

Blood contains many proteins (called clotting factors) that help stop bleeding. These clotting factors are numbered from 1 through 13, using roman numerals (such as I, IV, or X), and work together to insure that a clot is properly formed. People with hemophilia have either a low level of these clotting factors in their blood or none at all; the lower the level of clotting factor – the more serious the hemophilia. The result: problems, such as bleeding, including internal bleeding into the joints and muscles, without an obvious cause and bleeding after an injury or surgery, are more likely.

There are several different types of hemophilia. The following two are the most common:

- Hemophilia A (Classic hemophilia). This type is caused by a lack or decrease of clotting factor VIII (8).
- Hemophilia B (Christmas disease). This type is caused by a lack or decrease of clotting factor IX (9).

What is Von Willebrand Disease (VWD)?

Von Willebrand Disease (VWD) is a disorder that is caused by a problem with one of the proteins in the blood (von Willebrand factor or VWF). People with VWD either don't have enough VWF or what they have doesn't work correctly.

VWF plays two important roles in blood clotting. It makes platelets stick to the wall of an injured blood vessel and to each other. Without it, a platelet plug cannot be made. VWF is also a called a carrier protein because it also carries one of the clotting factors, factor VIII (8), with it. This means that it helps make sure there is enough factor VIII (8) is in the blood that it gets to where it's needed. Without VWF, factor VIII (8) will be broken down in the bloodstream and there may not be enough of it to help make a firm clot.

There are six different types of VWD:

- **Type 1:** This is the most common and mildest form of VWD. A person with Type 1 VWD has lower-than-normal levels of VWF and may also have low levels of factor VIII (8), which is another type of blood-clotting protein.
- **Type 2:** (with subtypes 2A, 2B, 2M and 2N): With this type of VWD, the body makes normal amounts of VWF but it does not work the way it should. Type 2 VWD is further broken down into four subtypes—2A, 2B, 2M, and 2N—depending on the specific problem with the person's VWF. Because each type is treated differently, a person should know which type he or she has.
  - *Subtype 2A* – Level of VWF is reduced, as is the ability of platelets to clump together.
  - *Subtype 2B* – Although the factor itself is defective, the ability of platelets to clump together is actually increased.
  - *Subtype 2M* – VWF levels are decreased and the interaction of VWF with platelets or connecting tissue is reduced.
  - *Subtype 2N* – The binding of VWF to factor VIII (8) is markedly decreased.
- **Type 3:** This is the most severe form of VWD, in which a person has very little or no VWF and low levels of factor VIII.

STEPS for LIVING
Education for all life stages.

Basics of Bleeding Disorders

How Does Someone Get a Bleeding Disorder?

While there are many different and rare bleeding disorders, generally bleeding disorders are inherited genetically. It is possible to develop a form of Von Willebrand Disease and hemophilia later in life as a result of other medical conditions (called Acquired Von Willebrand Syndrome and Acquired Hemophilia respectively) and about one-third of hemophilia patients have hemophilia not as a result of their parents genetically passing it on, but due to a change (called spontaneous mutation) in the gene's instructions for making the clotting factor protein. This change can prevent the clotting protein from working properly or the protein may be missing altogether.

Hemophilia is caused by a problem in one of the genes that tells the body to make the clotting factor proteins needed to form a blood clot. These genes are located on the X chromosome. All males have one X and one Y chromosome (XY) and all females have two X chromosomes (XX).

Males who inherit an X chromosome that has the abnormal gene on it have hemophilia. Rarely, a condition called "female hemophilia" occurs. In these cases, both X chromosomes are affected or one of two X chromosomes is missing or inactive. In these women, bleeding symptoms may be similar to those that hemophilia men have.

A female who inherits one affected X chromosome is said to become a carrier of hemophilia. In other words, she "carries" the gene that causes hemophilia on a chromosome. These women sometimes have low factor levels as well and can have symptoms of hemophilia. In addition, they can pass the affected gene on to their children. There will be a 50% chance their sons will have the bleeding disorder and a 50% chance their daughters will be carriers.

Hemophilia is a one of the more common inherited bleeding disorders. It occurs in about 1 of every 5,000 male births. Currently, about 20,000 men and boys in the United States have hemophilia. (Although hemophilia can occur in women, the number of women affected with hemophilia is extremely small.) Hemophilia affects people from all racial and ethnic groups.

The two most common forms of hemophilia are hemophilia A and hemophilia B.
Hemophilia A is about four times more common than hemophilia B, and about half of those with hemophilia A have a severe form. That means they have less than 1% of factor VIII, the clotting factor they need.

About 10%–15% of people with hemophilia develop an antibody (called inhibitor) to the clotting factors used to treat the bleeding disorder. An antibody is the body's natural response to fight off a "foreign" invader, such as a virus. When people develop an inhibitor, their body sees the clotting factor as a foreign body and fight against it, and clotting factor does not stop the bleeding.

For people who develop inhibitors, treating a bleed becomes extremely difficult, and the cost of care can increase because more clotting factor or a different type of clotting factor is needed.

STEPS for LIVING
Education for all life stages.

Basics of Bleeding Disorders

Patients with inhibitors often experience increased joint disease and other complications from bleeding, which reduce quality of life.

Von Willebrand Disease is passed genetically from parents to children. Types 1 and 2 VWD can develop when only one parent carries the gene. Type 3 VWD is passed along only if both parents pass the gene to the child.

Treatment for Bleeding Disorders

In general, small cuts and scrapes are treated with usual first-aid measures: clean the cut and then apply pressure and a bandage. Deep cuts or internal bleeding, such as bleeding in to the joints or muscles, require treatment by replacing the missing clotting factor to reach levels close to normal, in order to produce a firm clot and stop the bleeding. These treatments are given through a needle into a vein (infusion), orally or by a nasal spray.

Clotting Factor: All replacement clotting factor treatments are infused. This means they are injected from a needle placed into the student's vein (referred to as intravenously). This process takes a little time; it isn't like getting a quick shot. Some students with limited venous access may use alternative access devices, such as a port or catheter that can be surgically inserted under the skin in the chest area to make it easier to administer clotting factor products.

- *Clotting factor concentrate* (called *factor*) is a dried powder form of the missing clotting factor; it is mixed with sterile water to become a liquid again before it is given. If the student has severe hemophilia, doctors may recommend giving clotting factor several times a week to prevent most bleeds (called prophylaxis). Prophylaxis reduces the number of bleeds but does not prevent all bleeds.

Desmopressin Acetate (DDAVP): A synthetic form of the naturally occurring hormone that can be given through a vein or by nasal spray. It helps release von Willebrand Factor (vWF) and factor VIII (8) from where it is stored in the body tissues. For people with mild, and some cases of moderate, hemophilia, and Von Willebrand Disease (VWD), it can help increase their own factor VIII (8) levels so that they do not have to use clotting factor.

Epsilon Amino Caproic Acid: Given either in a vein or taken by mouth (as a pill or a liquid) this is a chemical that prevents clots from breaking down, which results in a firmer clot. It is often used for bleeding in the mouth or after a tooth has been removed because it blocks an enzyme in the saliva that causes clots to come apart.

Medical Emergencies for Bleeding Disorders

Prevent bleeding as soon as possible; treat immediately when it occurs - Depending on the severity of the underlying bleeding disorder, bleeding episodes may be frequent to rare. When possible, prevention of bleeding is the goal in managing patients with a bleeding disorder. However, when bleeding does occur, it is important to recognize the signs and symptoms at the earliest possible time and treat it appropriately.

STEPS for LIVING
Education for all life stages.

Basics of Bleeding Disorders

Some bleeds are more serious than others - Common bleeds, such as bruises, nosebleeds, mouth bleeds and heavy periods generally do not require extraordinary emergency medical attention. If a person with a bleeding disorder has a bleed and is not on a home-therapy program that person should contact a doctor or hemophilia treatment center (HTC) to determine what type of treatment is required.

Potential bleed site are:

- Mouth Bleeds
- Urinary Tract Bleeds
- Nose Bleeds
- Joint Bleeds
- Muscle Bleeds
- Soft Tissue Bleeds
- Head Bleeds **(911!*)**
- Eye Bleeds **(911!*)**
- Spinal Cord **(911!*)**
- Neck or Throat Bleeds **(911!*)**
- Deep Cuts or Lacerations **(911!*)**
- Gastrointestinal (GI) Bleeds **(911!*)**

911! Bleeding from these sites is life threatening. If a person has bleeding at one of these sites, call 911 immediately, notify the person's emergency contact and, if protocol indicates, also the hemophilia treatment center (HTC), and bring the person's treatment (factor) with them to the emergency room.

Preventive measures to support people with bleeding disorders

Physical Activity: Physical activity is any activity that causes your body to work harder than normal. Such activities are beyond the daily routine of sitting, standing, and walking up stairs. Everyone can benefit from increased physical activity. Some physical activities are considered safer than others for people with bleeding disorders. Avoid contact sports and other forms of hard, physical exercise involving strenuous exertion that could precipitate a bleed.

Encourage moderate exercise. Listen to the individual who will come to know their own safe limits of physical activity. The IHP should address which activities are suggested for this individual student.

Do not mock requests if a person with a bleeding disorder must stop participating in an activity because of pain or a bleed. It is important to encourage behavior that will maintain the individual's health.

Pain: When a person has a bleed it can be quite painful. The individual should have a plan created with input from his or her hemophilia treatment center or hemophilia specialist. It is very important that the plan includes instructions about how to handle a painful bleed.

Adaptive Devices: From time to time the individual may need to use adaptive devices such as crutches or a wheel chair. Do not draw undue attention to the person while at the same time making sure that ample exceptions are afforded the individual, as needed.

STEPS for LIVING
Education for all life stages.

Basics of Bleeding Disorders

Treatment: An important aspect of maintaining a health body is creation of a treatment plan which should include arrangements for administering treatment. Treating early is the key with bleeding disorders to avoid continued bleeding and further damage.

Awareness: Each individual may have a particular desire regarding if, w ho and how much to tell about his or her bleeding disorder. For younger individuals in a school setting all staff that have regular contact with the student should be made aware of their bleeding disorder. Staff needs to know what to do if the student has a bleed and needs to learn to listen to the student if he or she is feeling unwell.

Abuse: Bruises on children or young adults can easily be misinterpreted as a sign of physical abuse. Even trained healthcare providers may have difficulty determining whether the bruising they see is associated with abuse or a medical condition, such as a bleeding disorder. Having a bleeding disorder does not exempt a child from abuse. If you suspect abuse, authority figures should look for specific bruising patterns such as slap marks or the outline of an object, such as a paddle or belt. A thorough investigation should occur before any accusations are made.

Additional Information and resources available at:
www.stepsforliving.hemophilia.org

OTHER RESOURCES
Center for Disease Control and Prevention
National Center on Birth Defects and Developmental Disabilities, Blood Disorders page
http://www.cdc.gov/ncbddd/blooddisorders/index.html