BLEEDING DISORDERS

Caring for Children with Bleeding Disorders in a Community Program

Unified Referral and Intake System (URIS)
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Manitoba

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INTRODUCTION

Unified Referral and Intake System

The Unified Referral and Intake System (URIS) is a joint collaboration of various government departments, health service organizations, school divisions, and child caring organizations. URIS supports community programs in the care of children with specific health care needs. Community programs that are eligible for URIS support include schools, licensed child care facilities and agencies providing respite services. URIS provides a standard means of classifying the complexity of health care needs and establishes the level of qualification required by personnel to support children with these health care needs. Health care needs that are classified as 'Group B' can be delegated to non-health care personnel who receive training and monitoring by a registered nurse. For children with 'Group B' health need s (e.g., bleeding disorder), the nurse provides the following support:

- develops and maintains a written health care plan;
- provides training to community program personnel that are responsible for the child; and
- monitors community program personnel that receive training.

This document provides standard clinical information that is relevant to the care of children with bleeding disorders in community program settings. Supplemental documents are also provided to assist the nurse in the development of health care plans and training and monitoring of community program personnel.

CLINICAL INFORMATION

The following information is considered 'best practice' in community program settings and is the basis for all bleeding disorder information contained in this document and its supplements.

Circulation System

Blood is carried throughout the body within a network of blood vessels. When a blood vessel is damaged, the blood leaks through the holes in the vessel wall. The vessels can break near the skin's surface, as in a cut or they can break deep inside the body, forming a bruise or an internal bleed (hemorrhage).

Platelets are small cells circulating in the blood that play an important role in stopping bleeding. Platelets clump together and form a plug. Then clotting proteins (e.g., Factor VIII and IX) in the blood stick to the platelets to form a clot.

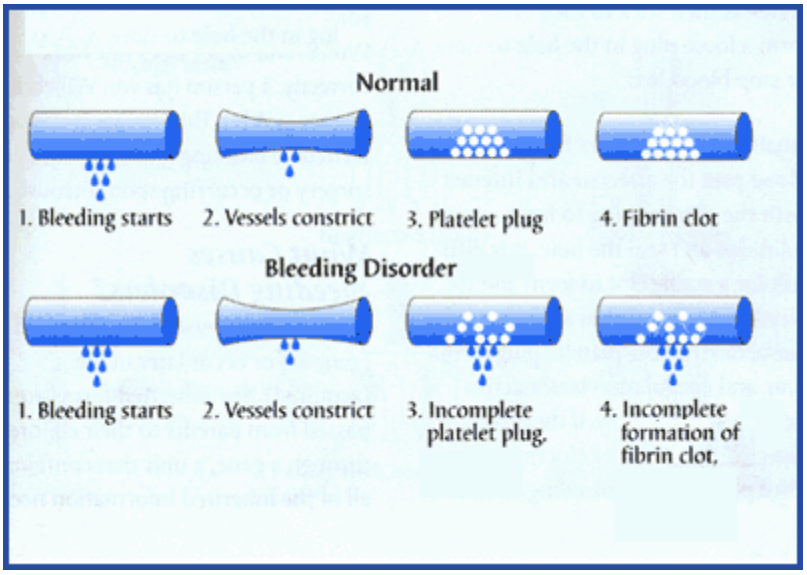

Figure 1

Stage 1: The blood vessel is damaged and the bleedings starts.

Stage 2: The blood vessel constricts to slow the flow of blood to the injured area.

Stage 3: Platelets stick to, and spread on, the walls of damaged blood vessel.

Stage 4: Clotting proteins circulating in the blood are activated on the surface of the platelets to form a clot. There are many proteins in the blood (e.g., Factor VIII, Factor IX, Von Willebrand Factor) that work together in a chain reaction. When one of the proteins is absent, the chain reaction is broken and clotting does not happen or it happens much more slowly.

Types of bleeding disorders

There are several different types of bleeding disorders which can vary in severity.

Hemophilia

When a person has hemophilia, one of the clotting proteins (Factor VIII or IX) is lacking and therefore their blood does not clot normally. A person with hemophilia does not bleed faster but can bleed for a longer time.

For a person with hemophilia, bleeding from a minor cut is usually not serious. Internal bleeding in joints, tissues and muscles is more serious. If bleeding occurs in a vital organ, especially the brain, it can be life-threatening. Brain hemorrhages are the leading cause of death from bleeding in hemophilia.

Until the age of 2, most bleeds are surface bruises. When babies are learning to walk, they fall frequently and suffer many bumps and bruises. Bleeding into the joints, soft tissues and muscles is seen more frequently after the age of two. Internal bleeding is often caused by minor injury - a bump or a slight twist of a joint. However, internal bleeding, especially among severe hemophiliacs, can happen for no apparent reason. In many children, symptoms become less severe as children move into adolescence and young adulthood, not because their hemophilia is less serious but they learn to avoid situations that might lead to bleeding.

Hemophilia is a hereditary condition, usually affecting males. It is a sex-linked genetic disorder as the genes responsible for producing Factor VIII and IX are situated on the X chromosome.

Hemophilia can also occur even if there is no family history. Many of these cases are the result of genetic mutations that occur at the time of conception.

Types of hemophilia

There are two types of Hemophilia – A and B.

Hemophilia A affects less than 1 in 10,000 people. It is also called Classic Hemophilia (it is the most common form of Hemophilia) or Factor VIII Deficiency (this is the clotting protein that is lacking).

Hemophilia B affects approximately 1 in 50,000 people. It is also called Christmas Disease (named after Steven Christmas, a Canadian, who was the first person to be diagnosed with Hemophilia B) or Factor IX Deficiency (this is the clotting protein that is lacking).
Hemophilia has three levels of severity including mild, moderate and severe. Although the severity of bleeding may differ for each person, they all have the potential to bleed when injured. A person with mild hemophilia has 5-30% of the normal level of Factor VIII or IX (which is normally between 50 and 150%) and has very few hemorrhages.

They may be aware of their bleeding problem only in the case of surgery, a tooth extraction or a serious injury. Women with mild hemophilia may bleed more during their menstrual period. A person with moderate hemophilia has 1-5% of the normal level of Factor VIII or IX. Their hemorrhages are often the result of minor trauma, such as a sports injury. A person with severe hemophilia has less than 1% of the normal level of Factor VIII or IX. They have hemorrhages several times a month and often there is no obvious cause for the bleeding.

Von Willebrand Disease (VWD)

Von Willebrand Disease (VWD) is the most common inherited bleeding disorder, affecting 1% of the population. There are different types of VWD but all are caused by a problem with Von Willebrand Factor (VWF), a clotting protein in the blood. The VWF acts as a glue to hold the platelets to the wall of the damaged vessel and to each other. When there is not enough VWF in the blood, or when it does not work properly, the platelets do not stick and it takes the blood longer to clot. The VWF also carries Factor VIII in the bloodstream. So when the VWF level is low, so is Factor VIII.

Von Willebrand Disease is a hereditary disorder. It affects men and women in equal numbers. If one or both of the parents have VWD, they can pass it on to their children. VWD can also occur if there is no family history. Many of these cases are the result of genetic mutations that occur at the time of conception

Types of VWD

Von Willebrand Disease is divided into three categories - Type 1, Type 2 and Type 3. Each type can vary in severity.

Type 1 VWD is the most common form, affecting 75% of all people with VWD. With type 1 VWD, the Von Willebrand Factor (VWF) works normally, but there is not enough of it. Many people have no symptoms of VWD at all until a bad injury or surgery at which time they could have serious bleeding. Other people have mild symptoms such as bleeding from their gums, minor bruises and prolonged bleeding from cuts. Some women with Type 1 VWD have heavy, prolonged bleeding during their periods. Injuries and surgery can lead to severe bleeding even in this mild form of VWD.

Type 2 VWD is much less common, representing 20-25% of all cases. In Type 2 VWD the amount of VWF in people's blood is often normal but it does not work properly. There are several sub-types of Type 2 VWD.

Type 3 VWD is very rare, affecting about 1 in 500,000 people. However, it is the most severe type of VWD, and resembles severe Hemophilia A. People with Type 3 VWD have very little VWF in their blood. As a result, bleeding happens often and, if untreated, can be serious.

Idiopathic Thrombocytopenic Purpura (ITP)

ITP is a disorder of the blood that involves the immune system. Platelets control and stop bleeding by causing blood to clot. In ITP, a specific type of antibody is produced

which coats the platelets and causes them to be destroyed. This can results in bleeding into the skin, mouth, noise and occasionally internally.

The cause of ITP is unknown. It can occur after a cold or viral illness, mumps, measles or chicken pox. It can also happen after taking some medicines. The majority of ITP cases in children are temporary. 85-90% of children with ITP recover within one year.

Treatment

There is no cure for hemophilia or Von Willebrand Disease. The disease is managed by prevention and should a bleed occur, prompt first aid should be initiated, and factor replacement may be required to stop the bleeding. First aid may be all that is needed for bleeds such as minor cuts, bruises or nose bleeds. However, bleeding into a joint or a muscle is never minor and specific treatment is essential. The type of treatment depends in part on the type of bleeding disorder.

Factor replacement therapy is used to treat hemophilia and VWD. A pharmacological preparation of Factor VIII, Factor IX or von Willebrand factor is injected intravenously to enable the body to form a clot at the injured site. For most situations, these factor concentrates are very effective in stopping bleeding. However, the injected clotting factors only stay in the body for a very limited time. Factor replacement therapy can be given on-demand (after the beginning of a bleed to stop the bleeding) or prophylactic ally (several times a week to prevent bleeding). Children with hemophilia and Von Willebrand's have a FactorFirst card that includes recommended factor replacement treatment. A copy of the Factor First card should be available at the community program so it can be provided to medical personnel in the event of an emergency.

People with severe hemophilia and children who are very active are more likely to receive factor concentrates.

Other treatments that may be used include Desmopressin (DDAVP) or Cyklokapron (tranexamic acid). Desmopressin is used for Type 1 Von Willebrand Disease and many patients with mild Hemophilia A. It works by releasing Von Willebrand Factor that is stored in the lining of the blood vessels. This helps in transporting factor VIII into the bloodstream. Cyklokapron (tranexamic acid) helps to maintain a more stable clot once it has formed by stopping the activity of an enzyme (called plasmin) that dissolves blood clots. .

The treatment for ITP varies. If a child has no bleeding, there is often no treatment. If treatment is required, it includes drugs that suppress the child's immune system including steroids and gamma globulin (IVIG). These treatments do not cure ITP but are used to keep the platelet count in a safe range. Steroids can cause mood changes, weight gain, puffiness in the face and neck and irritation of the stomach. IVIG may cause temporary headaches, nausea, lightheadedness or slight fever. In very serious cases where medication does not work, a splenectomy (removal of the spleen) may be recommended.

Precautions

A child with a bleeding disorder should be encouraged to participate in physical activities that keep his/her muscles and joints strong. Being in good physical condition can actually reduce the number of bleeding episodes a person has.

Many people with mild forms of a bleeding disorder can participate in all kinds of sports including active sports like soccer and high-risk sports like skiing. People with more severe forms of a bleeding disorder may find these activities lead to serious bleeding and high-risk activities are strongly discouraged.

It is recommended to talk to the parent/guardian if a child wants to participate in higher risk activities such as football, wrestling, ice hockey, full contact soccer or lacrosse, downhill skiing, boxing or rugby.

The following medications can affect the platelets ability to plug holes in blood vessels and should not be given to a person with a bleeding disorder.

- aspirin (e.g., Alka-Seltzer, Anacin, Aspirin, Bufferin, Dristan, Midol, 222)
- non-steroidal anti-inflammatory drugs (e.g., indomethacin and naproxen, Ibuprofen, Advil, Motrin);
- blood thinners (e.g., warfarin, heparin)

Acetaminophen (e.g., Tylenol) can be used for fever, headaches and minor aches and pains.

It is recommended that a person with a bleeding disorder wear Medic-Alert® identification.

External bleeds

For a person with a bleeding disorder, most bleeds are routine and emergencies are rare.

Surface cuts

Cuts and scrapes are treated the same as for any child.

1. Put on protective gloves.
2. Clean skin.
3. Apply firm continuous pressure until bleeding stops.
4. Apply a band-aid or dressing.
5. Encourage ice.
6. Call parent/guardian for instructions if bleeding has not stopped after 20 minutes. If unable to contact parent/guardian or emergency contact, call 911/EMS.
7. Provide medical personnel with copy of FactorFirst card, if available.

Nose bleeds

Nose bleeds may vary in children and are usually not serious. A nose bleed is treated the same as for any child.

1. Put on protective gloves.
2. Encourage the child to gently blow his/her nose to remove mucous and unstable clots. Once bleeding has stopped, encourage the child to NOT blow his/her nose again for as long as possible (at least 1-2 hours)
3. Position the child sitting with head slightly forward.
4. Apply firm continuous pressure for a minimum of 10 minutes or until bleeding stops. Use a cold cloth if possible.
5. Call parent/guardian for instructions if bleeding has not stopped after 20 minutes. If unable to contact parent/guardian or emergency contact, call 911/EMS.
6. Provide medical personnel with copy of FactorFirst card, if available.

Mouth bleeds

Bleeding in the mouth may be messy but is usually minor. Blood mixed with saliva can make the bleeding look worse than it is. Encourage the child to spit out rather than swallowing the blood as it can upset their stomach.

1. Put on protective gloves.
2. Apply firm continuous pressure (when applicable) until bleeding stops. Use a cold cloth if possible.
3. Encourage popsicles or ice.
4. Call parent/guardian for instructions if bleeding has not stopped after 20 minutes. If unable to contact parent/guardian or emergency contact, call 911/EMS.
5. Provide medical personnel with copy of FactorFirst card, if available.

Joint and muscle bleeds

It may not be obvious when bleeding starts in a joint or muscle. The child may be reluctant to use a limb or it may feel tight. It will gradually swell, feel hot to touch and become painful. Bleeding most commonly occurs in ankles, knees and elbows. Muscle bleeds can occur anywhere in the body.

1. Have the child rest. Keep the child still to avoid further injury.
2. Apply ice to injury. Do not leave it longer than 20 minutes and keep a cloth layer between ice and bare skin.
3. Elevate the injury body part.
4. Contact parent/guardian. If unable to contact parent/guardian or emergency contact after 15 minutes, call 911/EMS.
5. Provide medical personnel with copy of FactorFirst card, if available.

Bruising
Children with hemophilia often have visible bruising which is usually not serious. Small bruises usually disappear on their own. If a child complains of ongoing pain at a bruised site, inform the parent/guardian.

Internal Bleeds

Bleeding into the head, eye, neck, chest or abdomen may be life-threatening and requires immediate medical attention.

Head injury
All head injures must be considered serious as there is a risk of a brain hemorrhage.

Possible signs of internal bleed

- Loss of consciousness
- Drowsiness
- Dizziness
- Irritability
- Lethargy
- Nausea and/or vomiting
- Dilated or unequal pupils
- Headache
- Confusion
- Unsteady gait

Injury to eye and/or surrounding area
An injury to the eye and/or surrounding area could potentially result in an eye-threatening injury (e.g., loss of vision).

Possible signs of internal bleed

- Pain
- Swelling

Neck injury
Neck bleeding is serious because of the potential to block the airway. Any injury to the neck area, as well as a child's expressing pain and tenderness should be attended to quickly.

Possible signs of internal bleed

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

Chest injury

Injuries to the chest may be very painful if there is bleeding into the muscles between the ribs.

Possible signs of internal bleed

- Pain in chest
- Difficulty breathing
- Coughing up blood
- Pale skin
- Lack of energy

Abdominal injury

A blow to the stomach is serious because of the potential injury to vital internal organs.

Possible signs of internal bleed

- Pain in abdomen or lower back
- Nausea or vomiting
- Blood in urine
- Black or bloody stool

Responding to internal bleeds

If a child suffers a significant injury to the head, eye, neck, chest or abdomen, with or without showing signs, call 911/EMS.

1. Call 911/EMS.
2. Do not move the child to prevent further injury, unless child is in an unsafe place.
3. Notify parent/guardian.
4. Provide medical personnel with copy of FactorFirst card, if available.

It is recommended that the child is transported to Children's Hospital or a facility that has factor replacement treatment available.

HEALTH CARE PLAN

When a community program receives URIS Group B support for children with URIS 'Group B' health care needs, a written health care plan is developed and maintained by a registered nurse on an annual basis, minimally. The development and implementation of the health care plan should reflect the principles of inclusion, normalization and independence. From a practical standpoint, these principles mean:

- A child with a bleeding disorder is foremost a child within a family, child-care facility, classroom or other community program.
- The environment should be changed to support the child, not the child changed to suit the environment.
- Interventions should be as non-intrusive as possible and be delivered in a manner that respects the child's dignity and privacy as well as the normal routines and patterns of the community program.
- The parent/guardian and child have rights and obligations and should be actively encouraged to participate in decisions affecting themselves and their children.

Consultation with the parent/guardian and community program is required to develop a health care plan that is relevant to the child's needs and the community program setting. For some children, the management of their bleeding disorder within the community program may be complex and require consultation with health care professionals who are involved in the management of the child's bleeding disorder.

When a child has multiple health care needs, all relevant information should be integrated into one comprehensive plan. The format of the health care plan should be user friendly and provide the community program with the information required to safely manage the child's health care needs.

The health care plan should be kept in a location that is both secure and accessible at the community program. All community program personnel that may be responsible for a child with a bleeding disorder should be aware of the location of the health care plan. It should also accompany the child on excursions outside the facility.

Content

The following information is included in the bleeding disorder health care plan. The *Bleeding Disorder Health Care Plan(s)* contains this information and is included as a supplement to this document.

Demographic information

- Child name
- Gender
- Birth date
- Community program name
- Parent/guardian name and phone number(s)
- Alternate emergency contact name and phone number(s)

- Physician(s)
  - Hematologist and phone number
  - Family physician/pediatrician and phone number

Medical information

- Medical diagnoses and other relevant conditions
- Known allergies
- Availability of Medic-Alert® identification
- Prescribed medications
  - If medication is administered at home, name of medication is required only.
  - If medication is administered at the community program, additional information is required. See *When medication is administered at the community program* (below) for more details.

Bleeding disorder information

- Type of bleeding disorder
- History
  - First diagnosed
  - History of bleeds
- Precautions, if applicable
- Activity restrictions, if applicable
- Copy of FactorFirst card – it is the responsibility of the parent/guardian to provide the community program with a copy of the FactorFirst card

It is recommended that the child is transported to Children's Hospital or a facility that has factor replacement treatment available.

Responding to bleeds

- External bleeds
  - Description of external bleeds (e.g., mouth bleed, nose bleed, surface cut, bruising, joint and muscle bleed)
  - Responding to external bleeds
- Internal bleeds
  - Symptoms of internal bleeds (e.g., head injury, chest injury, abdominal injury, neck injury)
  - Responding to internal bleeds

Documentation

- Template for recording interventions and actions performed by nurse and/or community program personnel (e.g., communication, actions taken)
- Signatures & dates
    - Nurse signature & date(s) of health care plan development/review
    - Parent/guardian signature & date

When medication is administered at the community program

When medication is administered to the child during attendance at the community program, the health care plan also includes the following information. The *Administration of Medication* contains this information and is included as a supplement to this document.

- Name of medication
- Dosage
- Time of administration
- Route of administration
- Location of medication at community program
- Steps in administering medication
- Written record for administration of medication
    - Date/time of administration
    - Initial/signature of community program personnel that administered medication
    - If administration not complete, indicate reason

TRAINING

When a community program receives URIS Group B support, training is provided to community program personnel by a registered nurse. Training is provided on an annual basis, minimally. The training of community program personnel should reflect the principles of adult learning. From a practical standpoint, these principles mean:

- Identifying and integrating the learning needs of participants into the training session.
- Information should be applicable to the participants' responsibilities and focus on what is most useful to them.
- Adults have accumulated a foundation of life experiences and knowledge and need to connect learning to this knowledge/experience base.
- An organized training session with clearly defined elements assists participants in identifying and attaining learning goals.

It is recommended that all community program personnel that may be responsible for a child with a bleeding disorder attend the training session. As an example, community program personnel that may be responsible for a child with a bleeding disorder may include:

- in schools - teachers, teaching assistants, school administrators, office staff, substitute teachers, bus drivers, lunch room supervisors;
- in licensed child care facilities - child care providers, child care directors; and
- in recreational programs – staff members, administrators, volunteers.

The community program is responsible to ensure personnel that may be responsible for a child with a bleeding disorder attend the training session. It is recommended to keep a written record that indicates community program personnel in attendance and date that training occurred.

Adequate time should be scheduled for training to ensure community program personnel obtain the knowledge and skill necessary to safely respond to the needs of children with a bleeding disorder in their facility. The amount of time required to train community program personnel will vary depending on several factors such as the existing knowledge of community program personnel, number of personnel attending the session and format of training resources used (e.g., PowerPoint, Worksheet).

Whenever possible, training should be scheduled when all community program personnel can attend to ensure service is provided in an efficient manner. If the training session is poorly attended (i.e., there is not an adequate number of community program personnel to safely address the child's anaphylaxis), additional training should be scheduled. If subsequent training sessions are also poorly attended, alternate strategies should be discussed with the community program to ensure training is provided in an efficient manner.

When the community program has not received training, a child with a bleeding disorder may attend the community program. In such situations, the community program's internal policy for emergency situations (e.g., call 911/EMS) is implemented, is required.

Content

The following clinical information and child specific information is included in the training session.

Standard clinical information

- Circulation system
- Types of bleeding disorders
- Treatment of bleeding disorders
- Precautions and restrictions
- Description of bleeding disorder
- External bleeds - description and how to respond
- Internal bleeds - description and how to respond

Child specific information

- Type of bleeding disorder
- History
  - First diagnosed
  - History of bleeds
- Precautions and restrictions, if applicable
- Availability of FactorFirst card
- Actions to implement for external and internal bleeds, if different from standard clinical information

Training Resources

The following resources are included as supplements to this document. If alternate resources are used, it is the responsibility of the nurse to ensure its content is consistent with the clinical information included in this document.

- *Bleeding Disorder Handout*
- *Bleeding Disorder PowerPoint*
- *Bleeding Disorder Worksheet* (Word and PowerPoint version) is recommended for community program personnel that have previously attended a bleeding disorder training session. The Microsoft Word version may be better suited for individuals or small groups. The Microsoft PowerPoint version may be more suitable for large group settings.

MONITORING

Monitoring of trained community program personnel by a nurse is required to ensure that the knowledge and skill necessary to safely care for children with bleeding disorders has been acquired and/or retained. Monitoring is required on an annual basis, minimally.

The frequency and timing of monitoring is based on the professional judgment of the nurse as well as the complexity of information taught, maturational issues and the skill demonstrated by community program personnel. The following strategies may be used for monitoring purposes.

- Completion of the *Bleeding Disorder Training Session Evaluation Form* by community program personnel that attend training, included as a supplement to this manual;
- Asking community program personnel questions during the training session to assess their knowledge of seizure disorders. The *Bleeding Disorder Worksheet* is included as a supplement to this document.

REFERENCES

All About Von Willebrand Disease. Canadian Hemophilia Society, 2007. 2nd edition.

Hemophilia: What School Personnel Should Know. Canadian Association of Nurses in Hemophilia Care.

Home Treatment Guide for People with Bleeding Disorders, 2009. Canadian Association of Nurses in Hemophilia Care (Western Division).

Von Williebrand Disease: What School Personnel Should Know. Canadian Association of Nurses in Hemophilia Care.

RESOURCES

The following list includes resources that may be relevant to community programs in the care of children with bleeding disorders. The purpose of these agencies/organizations may not be consistent with the purpose and content of this manual.

Canadian Hemophilia Society
www.hemophila.ca

Hemophilia Society: Manitoba Chapter
www.hemophiliamb.ca

World Federation of Hemophilia
www.wfh.org

National Hemophilia Foundation
www.hemophilia.org