A School Nurse’s Guide to Bleeding Disorders:
Von Willebrand Disease & Hemophilia

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Target Audience

- Nurses in schools attended by a child with a bleeding disorder
- Teachers, physical education teachers, administrators, and guidance counselors
- Other healthcare professionals who require an introduction to school-related considerations for students with bleeding disorders
Objectives

- Identify signs and symptoms of VWD
- Recognize common treatments utilized for VWD within the school setting
- Describe the impact of a child with hemophilia in the school setting
- Integrate strategies in the school setting to promote a safe and positive learning environment
A School Nurse’s Guide to Bleeding Disorders:
Von Willebrand Disease Basics
What Is a Bleeding Disorder?

Students with bleeding disorders are missing or have ineffective clotting factors

- Students will bleed longer compared to those whose blood clotting factors are normal
- Bleeding disorders range from mild to severe

Common myths about students with bleeding disorders

- May bleed to death from superficial cuts
- Bleed faster
- Cannot participate in physical activities
- Have frequent school absences

How Does Bleeding Start and Stop?

http://wfh.org/2/docs/Publications/General_Guides/HIP_Educators-Guide.pdf
What is Von Willebrand Disease?

Caused by a deficiency or defect of the protein Von Willebrand factor (VWF) that is needed for blood to clot

- VWF is a protein that helps platelets adhere to one another and to the damaged area in the blood vessel
- Platelets are small cells in the body that plug areas of injury
- VWF also binds and protects clotting factor VIII in the circulation

Genetics & Incidence of VWD

- VWD is an inherited bleeding disorder
  - If a parent is affected, there is a 50% chance of passing the condition to each child
    - Multiple family members affected

- Most common genetically transmitted bleeding disorder
  - Incidence up to 1-3% of population (>3 million Americans)

- Individuals may experience symptoms for years prior to diagnosis

Patient/Family History

- Have you/your child or a blood relative ever needed medical attention for a bleeding problem or been told that you have a bleeding disorder?
- Any history of unexpected or excessive bleeding?
  - During/after surgery
  - With dental procedures, extractions
  - With trauma
  - During childbirth or heavy menstruation
  - Ever have bruises with lumps

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- National Heart Lung and Blood Institute (NHLBI). *Diagnosis, Evaluation, and Treatment of Von Willebrand Disease: U.S. Department of Health and Human Services (HHS); 2007*
Common Symptoms of VWD

- Abnormal Bruising
- Frequent Nosebleeds ≡ Epistaxis
- Excessive bleeding following dental extraction
- Excessive menstrual bleeding ≡ Menorrhagia
- Bleeding after injury or surgery
  - Removal of tonsils and adenoids is high risk

Epistaxis

- \(\frac{1}{3}\) of children that present with recurrent epistaxis have an underlying bleeding disorder
  - Positive family history & abnormal APTT useful predictive data

Frequency
- Times per day, week, month, or year
- Clustered

Severity
- Duration
- Associated Anemia

ENT evaluation
- Nasal Cautery

Case Study: 7 Year Old Male with Epistaxis

<table>
<thead>
<tr>
<th>Level of Concern</th>
<th>Low</th>
<th>High</th>
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<tbody>
<tr>
<td>Epistaxis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Frequency</td>
<td>Few times yearly</td>
<td>1-2 times monthly</td>
</tr>
<tr>
<td>Duration</td>
<td>~3 minutes</td>
<td>&gt;5 minutes</td>
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<tr>
<td>Seasonality</td>
<td>Yes</td>
<td>+ / -</td>
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<tr>
<td>Clusters</td>
<td>None</td>
<td>Yes</td>
</tr>
<tr>
<td>Interventions required</td>
<td>Pressure</td>
<td>More than pressure</td>
</tr>
</tbody>
</table>
Epistaxis

- The student should be in an upright position with their head tilted forward.
- Using universal precautions, apply firm, continuous pressure to the nose for 20 minutes.
- If bleeding continues, notify the parent, as treatment may be indicated.
  - If bleeding stops, student may return to classroom.
    - Always notify the parent if a bleeding episode occurs.
Normal menstrual cycle

- Cycle 21-35 days
- Duration 2-7 days
- Average blood loss 35-150ml total
- Average pad holds 5-15ml
- Average tampon holds 5ml

• Always™. “Your Period”. Available
Menorrhagia

- Prolonged and heavy menstrual bleeding is the most common complication reported by women and teenagers

- Students may need more time between classes or frequent restroom breaks to avoid accidents

- Goal of therapy is to prevent frequent school absences
  - Notify the hemophilia treatment center (HTC) if there are problems
Menorrhagia

- Defined as blood loss > 80 ml per cycle
- Questionnaire in 952 women with complaints of excessive menstrual bleeding
  - 226 / 952 participants consented to have blood loss measured
  - Median blood loss ~53 ml
  - 34% experienced blood loss > 80 ml

Predictors of menorrhagia
- Clots greater than 1 cm
- Iron deficiency
- Changing protection more frequently than hourly

76% prediction of > 80 ml ⇒ menorrhagia if above are present

Case Study: 16 Year Old Female

- Hospitalized: Profound anemia due to menorrhagia
- Menstrual history
  - Menarche 12 years
  - Regular, last 7-10 days, require ~10 pads daily
- Frequently missed school during menses due to concern of experiencing “accidents during class”
- Recent symptoms of worsening fatigue, SOB with activities
- Laboratory evaluation
  - Severe iron deficiency anemia
  - Decreased FVIII, VWF:Ag, VWF:RCo, abnormal multimers
Case Study: 16 Year Old Female

- GYN referral: Moderate anemia due to menorrhagia
- Hormonal therapy instituted with improved symptoms
  - Duration 2 months
- Family history
  - Maternal hysterectomy at 40 years due to menorrhagia
  - Brother with epistaxis
- Consider
  - Holding hormonal suppressive therapy & return in 2 months for evaluation → Not a good idea if she is still anemic
  - Evaluate other symptomatic family members → Level of suspicion increased if VWD diagnosed
  - Utilize strategies for treatment of VWD → monitor symptoms
Surgery & Invasive Procedures: Contributors to Level of Suspicion

- Tonsillectomy / adenoidectomy
  - Early or delayed bleeding when eschar sloughs
  - Incidence of hemorrhage in general population
    - ≤ 5 days: 45%
    - > 5 days: 55%

- Dental extractions
  - Abnormal bleeding including liver clots
- Requirement for nasal cautery or ENT intervention
- Need for blood transfusion related to surgical blood loss
Common Symptoms of VWD

With proper medical treatment, many of these symptoms can be eliminated/controlled.
Types of Von Willebrand Disease

- **Type 1**
  - Most common (80%)
  - Least severe
  - VWF quantity decreased

- **Type 2**
  - Less common
  - VWF structure abnormal

- **Type 3**
  - Least common
  - Most severe
  - VWF absent


http://www.hemophilia.org/NHFWeb/MainPgs/MainNHF.aspx?menuid=182&contentid=47rptname=bleeding
A School Nurse’s Guide to Bleeding Disorders: VWD Treatment
Treatment Plans

Treatment may be preventative or may be used for a bleeding event

Preventative

- Hormonal therapy to control menstrual cycles
- Nasal sprays to prevent nose bleeds
- Infusion of replacement of Von Willebrand factor; less common
- Treatment prior to surgery or dental procedure

Treating bleeding episodes

- At the start of the bleeding event
- Treatment may continue for a period of time to allow healing
Treatment: Desmopressin Acetate

Desmopressin acetate: synthetic derivative of naturally occurring hormone called vasopressin

- Works through release of Von Willebrand factor from storage sites

- Desmopressin Nasal (Stimate®)
  - Most commonly used formulation
  - Intranasal spray

- Desmopressin Oral (DDAVP®)

- Common side effects: flushing of face, change in blood pressure, headache and stomach upset

Treatment: Antifibrinolytic Therapy

- Tranexamic acid (Lysteda®)/Aminocaproic acid (Amicar®)
  - Commonly prescribed oral antifibrinolytics used to treat menorrhagia, epistaxis, or following surgery or dental work
  - Antifibrinolytics work by preventing the breakdown of the clot once formed
  - Common side effects: nausea, vomiting, diarrhea, abdominal pain and muscle cramps
  - Aminocaproic acid compounded into a nasal spray

Medical and Scientific Advisory Council (MASAC) recommendation #196
http://www.hemophilia.org/NHFWeb/MainPgs/MainNHF.aspx?menuid=1&contentid=29
Treatment: Factor Concentrate

Factor concentrate containing intact Von Willebrand factor

• Replaces defective or absent Von Willebrand factor

• Intravenous administration

• Usually given at treatment center or medical facility

• Students and families may be trained to administer intravenous infusions at home

Medical and Scientific Advisory Council (MASAC) recommendation #196
http://www.hemophilia.org/NHFWeb/MainPgs/MainNHF.aspx?menuid=1&contentid=29
Treating Pain

Avoid medications that increase the risk of bleeding
- Aspirin
- Long term use of nonsteroidal anti-inflammatory medications
  - ibuprofen, naproxen

Acetaminophen is a pain reliever that does not increase bleeding risk and is generally considered safe in students with bleeding disorders.
A School Nurse’s Guide to Bleeding Disorders: Hemophilia
Hemophilia Myths

- Bleed to death from a cut
- Decreased lifespan
- Cannot participate in physical exercise
- Will have frequent absences due to hemophilia-related problems
Hemophilia affects approximately 1 in 5,000 males

Bleeding disorder caused by absence or decreased amount of clotting factor

Factor FVIII deficiency/Hemophilia A is the most common type

Factor IX deficiency/Hemophilia B (also known as Christmas disease) is less common

Is All Hemophilia the Same?

Degrees of Severity

- **Normal**
  - Factor VIII or IX activity
  - 50 - 150%

- **Mild**
  - Factor VIII or IX activity
  - 5 - 40%

- **Moderate**
  - Factor VIII or IX activity
  - 1 - 5%

- **Severe**
  - Factor VIII or IX activity
  - < 1%

Why Do People with Hemophilia Sometimes Bleed Longer than Other People?

Normal Clotting Process

Clotting in Hemophilia

Is Hemophilia Lifelong?

Hemophilia is an Inherited Disorder

- Hemophilia A and B are X-linked recessive disorders
- Affects all races and ethnic groups
- Typically expressed in males and carried by females
- Severity level and type is consistent between family members
- 30% of cases of hemophilia are new mutations (i.e., no known family history)

Hemophilia A (Factor VIII Deficiency). National Hemophilia Foundation (NHF), 2006.
A School Nurse’s Guide to Bleeding Disorders:
Bleeding Episodes
What Causes a Joint Bleed?
What Happens in a Joint Bleed?
Which Joint Bleeds are Most Common?

What Happens in a Muscle Bleed?
Which Muscle Bleeds are Most Common?
Which Bleeds are Serious or Life-Threatening?
Treatment of Bleeding Episodes

Why Should Bleeds be Treated Quickly?

Treating Bleeds with Factor Replacement Therapy

- **Episodic/On-Demand**
  - Factor administered after the bleed occurs

- **Prophylaxis**
  - Factor administered before the bleed occurs
  - Scheduled administration
Treating Bleeds with First Aid

R.I.C.E.
Rest
Ice
Compression
Elevation

How Can Bleeds be Managed in the School?

- Analgesics depending on pain severity and chronicity

- Learning environment accommodations as needed
  - P.E. class activities
  - Increased time to walk between classes
  - Use of elevator
  - Use of crutches

A School Nurse’s Guide to Bleeding Disorders: Physical Activity and Hemophilia
Exercise and Physical Activity are Important!
General Activity Guidelines for Schools to Consider

- Encourage participation in regular physical activity
- Infuse prophylactically if recommended by medical team
- Avoid physical activity during active bleeding
- After a bleeding episode, resume activity only when motion, strength, coordination restored

## What Activity Selection is Appropriate for the Student?

<table>
<thead>
<tr>
<th>Activity</th>
<th>Rating</th>
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<tbody>
<tr>
<td>Aerobics</td>
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<td>Archery</td>
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<td>Aquatics</td>
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<td>Bicycling</td>
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<td>BMX Racing</td>
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<td>Bowling</td>
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<td>Boxing</td>
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<td>Canoeing</td>
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<td>Cardiovascular Training Equipment</td>
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<td>Elliptical Machine</td>
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<tr>
<td>Rowing Machine</td>
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<tr>
<td>Ski Machine</td>
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<tr>
<td>Stationary Bike</td>
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<tr>
<td>Stepper</td>
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<tr>
<td>Treadmill</td>
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<tr>
<td>Cheerleading</td>
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<td>Circuit Training</td>
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<tr>
<td>Dance</td>
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<tr>
<td>Diving/Competitive</td>
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<tr>
<td>Diving/Recreational</td>
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<tr>
<td>Exercise Classes</td>
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<td>Body Sculpting</td>
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<tr>
<td>Cardio Kick-Boxing</td>
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<td>Physio Ball</td>
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<td>Spinning</td>
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<td>Fishing</td>
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<table>
<thead>
<tr>
<th>Activity</th>
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<tr>
<td>Football</td>
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<td>Frisbee</td>
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<td>Frisbee Golf</td>
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<tr>
<td>Ultimate Frisbee</td>
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<tr>
<td>Golf</td>
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<tr>
<td>Gymnastics</td>
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<tr>
<td>Hiking</td>
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</tr>
<tr>
<td>Hockey (Field, Ice, Street)</td>
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<tr>
<td>Horseback Riding</td>
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<tr>
<td>Ice-Skating</td>
<td>2.5</td>
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<tr>
<td>Inline Skating</td>
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</tr>
<tr>
<td>Jet Skiing</td>
<td>2</td>
</tr>
<tr>
<td>Jumping Rope</td>
<td>2</td>
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<tr>
<td>Kayaking</td>
<td>2.5</td>
</tr>
<tr>
<td>Lacrosse</td>
<td>2</td>
</tr>
<tr>
<td>Martial Arts - Karate/Kung Fu/Tae Kwon Do</td>
<td>2.5</td>
</tr>
<tr>
<td>Martial Arts/Tai Chi</td>
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<tr>
<td>Motorcycling/Motor Cross Racing</td>
<td>3</td>
</tr>
<tr>
<td>Mountain Biking</td>
<td>2.5</td>
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<tr>
<td>Pilates</td>
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<tr>
<td>Power Lifting</td>
<td>3</td>
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<tr>
<td>Racquetball</td>
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<tr>
<td>River Rafting</td>
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<tr>
<td>Rock Climbing (Indoor/Challenge Course)</td>
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<tr>
<td>Rock Climbing (Natural Setting)</td>
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</tr>
<tr>
<td>Rodeo</td>
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<tr>
<td>Roller-skating</td>
<td>2</td>
</tr>
<tr>
<td>Rowing/Crew</td>
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</table>

Category 1
- Most students with hemophilia can safely participate
- Can be enjoyed over a lifetime

Examples: Bicycling, golf, swimming and hiking
Category 2:
- The physical, social, and psychological benefits may outweigh the risk in these sports
- Majority of sports fall into this category

Examples: Baseball, soccer, gymnastics, skateboarding, bowling, basketball and skiing
Category 3:

- By nature, these activities are dangerous even for those without hemophilia
- The risks outweigh the benefit in these sports

Examples: Football, wrestling and hockey
A School Nurse’s Guide to Bleeding Disorders: Coordination of Care with the Indiana Hemophilia & Thrombosis Center

The Indiana Hemophilia & Thrombosis Center is committed to providing the highest quality comprehensive healthcare services to patients with bleeding and clotting disorders, and to their families.

8402 Harcourt Rd. Ste. 500
Indianapolis, IN 46260
1-877-256-8837
www.ihtc.org
The Comprehensive Care Team

IHTC uses a Comprehensive Care Model, providing access to multidisciplinary healthcare professionals:

- Hematologists
- Nurses
- Physical Therapists
- Social Workers
- Career Counselors
- Genetic Counselors
- Registered Dietitians
- Dental Hygienists

Hemophilia Treatment Centers (HTCs). Centers for Disease Control and Prevention (CDC).
http://www.cdc.gov/ncbdd/hemophilia/HTC.html
HTC Nurse

- Primary contact for creation of Individualized Health Plan
- Available by telephone for triage of bleeding episodes
- Resource for hemophilia education and guidance
What Is an Individualized Healthcare Plan?

- A written document that outlines the provision of student healthcare services
- Intended to achieve specific student outcomes
- Created for students with significant or chronic health problems
- Fulfill administrative and clinical purposes:
  - Management of health conditions to promote learning
  - Facilitation of communication, coordination, and continuity of care
  - Evaluation/revision of care provided

How is an IHP Developed?

- School nurse is responsible and accountable for creating the IHP, for managing its activities and for its outcomes\(^1\)

- Developed collaboratively
  - Family
  - Student
  - Student’s healthcare providers
  - School staff

- Indiana Hemophilia & Thrombosis Center can assist in formulation of the IHP

What Medical Information Should be Carried?
Developing a Plan for Emergency Contact and Care

- Primary contact = Parents / Guardian

- Secondary contact = Hemophilia Treatment Center

- St. Vincent Indianapolis Hospital versus local hospital
  - Highly specialized nature of hemophilia care
  - Availability of resources
HTC Career Counselor

- Liaison between home and school
- Primary contact for Section 504 Plan and Individualized Education Plan (IEP) case conferences
- Available to assist students in meeting academic goals
Promoting a Safe and Positive Learning Environment
Review of Objectives

- Identify signs and symptoms of VWD
- Recognize common treatments utilized for VWD within the school setting
- Describe the impact of a child with hemophilia in the school setting
- Integrate strategies in the school setting to promote a safe and positive learning environment