

Bleeding Disorders in Women

What is going on down there?

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Hemophilia
of Georgia



It takes more than medicine...

Objectives

- Be able to recognize the signs and symptoms of a bleeding disorder in females.
- Discuss the current treatments available for a women with a bleeding disorder.
- List the most common bleeding disorders that affect females.
- Identify the challenges that are associated with females with a bleeding disorder.

Bleeding In Women

Question:

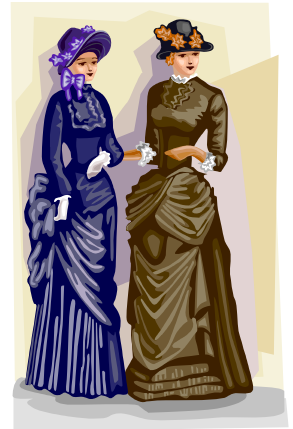
- What did women use for sanitary protection before commercial pads and tampons?



Bleeding In Women

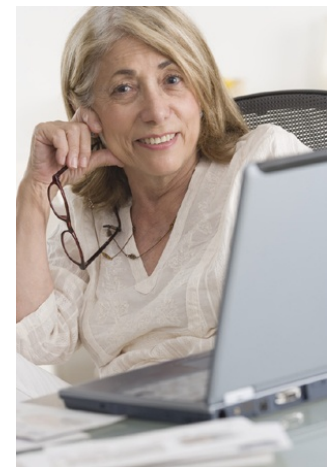
Answer:

- You might be surprised to learn that, according to the Museum of Menstruation, in the 19th century American and European women probably wore nothing to contain menstrual bleeding. In fact, not only did women wear nothing to absorb menstrual blood, they also did not wear underwear.



Bleeding Disorders in Women

- For women, an inherited bleeding disorder is particularly problematic due to the monthly onset of menstruation
- Excessively heavy menstrual bleeding: menorrhagia
 - Most common symptom of bleeding disorder
 - Common complaint



Public Health Relevance Menorrhagia

- Menorrhagia \geq 80mL blood loss per period
- A large proportion of women consult a physician during lifetime for heavy menstrual bleeding
 - 62/1000 annually
 - 2.9 million women
 - 4 million doctor visits



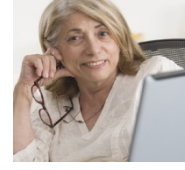
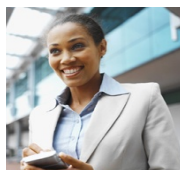
Public Health Relevance Menorrhagia

- Difficult to define
 - Cultural factors
 - Women's and physician's perceptions of what is a normal period
 - How to diagnose?
- In 50% of cases, no organic pathology determined and diagnosis is dysfunctional uterine bleeding.



Public Health Relevance Menorrhagia

- **Impact on women's lives**
 - Diminished quality of life, home, and work
 - Anemia, requiring medical treatment
 - Surgical procedures
 - 20% of hysterectomies performed for “dysfunctional uterine bleeding”
 - More than 25% of US female population undergo hysterectomy by age 60
 - Estimated annual cost of hysterectomies in US > \$5 billion

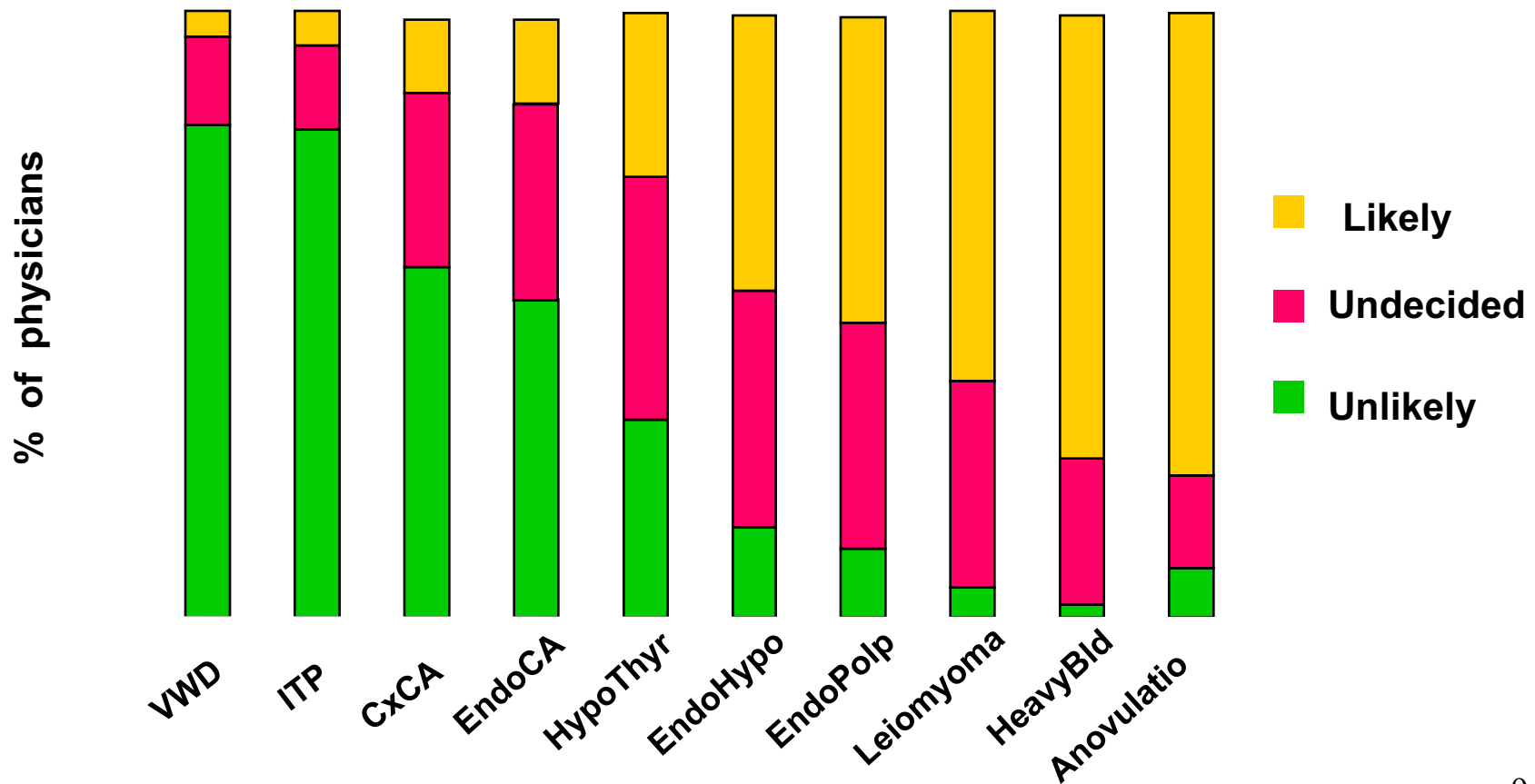


CDC Collaborative Research

Part I: Physician Survey

ACOG Members in Georgia – 54% Response

Perceptions of Certain Conditions as Causes of Menorrhagia Among Reproductive-Aged Women (15 to 44)



CDC Collaborative Research Part I: Physician Survey

- CDC survey of OB/GYNs
 - Perceived prevalence of bleeding disorders among women with menorrhagia <1%
 - 42% reported never having seen a menorrhagia patient with a bleeding disorder

J Womens Health & Gender-Based Medicine. 2002;11:39-44.

CDC Collaborative Research Part I: Physician Survey

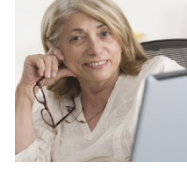
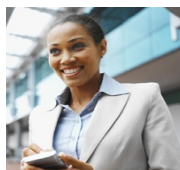
- Recent research indicates a higher prevalence than perceived
 - 1996–Sweden (Edlund, et al) 37%
 - 1998–England (Kadir, et al) 17%
 - 2004–Review of 11 studies (Shankar, et al) 11% to 15.6% for VWD
 - 2001–US (Dilley, et al)

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CDC Collaborative Research

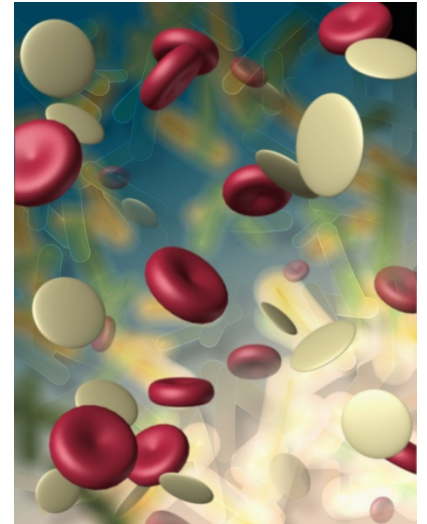
Part III: Management

- The population
 - 86% reported history of menorrhagia
 - It took an average of 16 years from onset of symptoms until diagnosis
 - More than half the women were tested multiple times before a diagnosis was made



How Bleeding Stops

- Vasoconstriction
- Platelet plug formation
- Clotting cascade activated
- Clotting factors are proteins in blood activated by platelets
- 12 factors cause fibrin to weave over the platelet plug and cause bleeding to stop



What is von Willebrand Disease?

- Hereditary bleeding disorder caused by the decreased production or decreased function of von Willebrand Factor (VWF)



Von Willebrand Disease

- Described in 1925 by Dr. Eric Von Willebrand
- Affects both males and females
- Most common inherited bleeding disorder
- Present in perhaps 1% to 3% of the population, over 100 times more prevalent than hemophilia

von Willebrand Disease

- Persons with vWD produce less VWF or produce a molecule that does not function normally
- When VWF is not present or does not function, platelets do not stick as readily to the damaged blood vessel

von Willebrand Disease

- Most people with this disorder have very mild symptoms
- Many of these people are undiagnosed unless a family member is identified or pre-surgical lab tests are done
- Some persons with this disorder can have life threatening bleeding episodes after surgery, childbirth, or extensive, traumatic injury

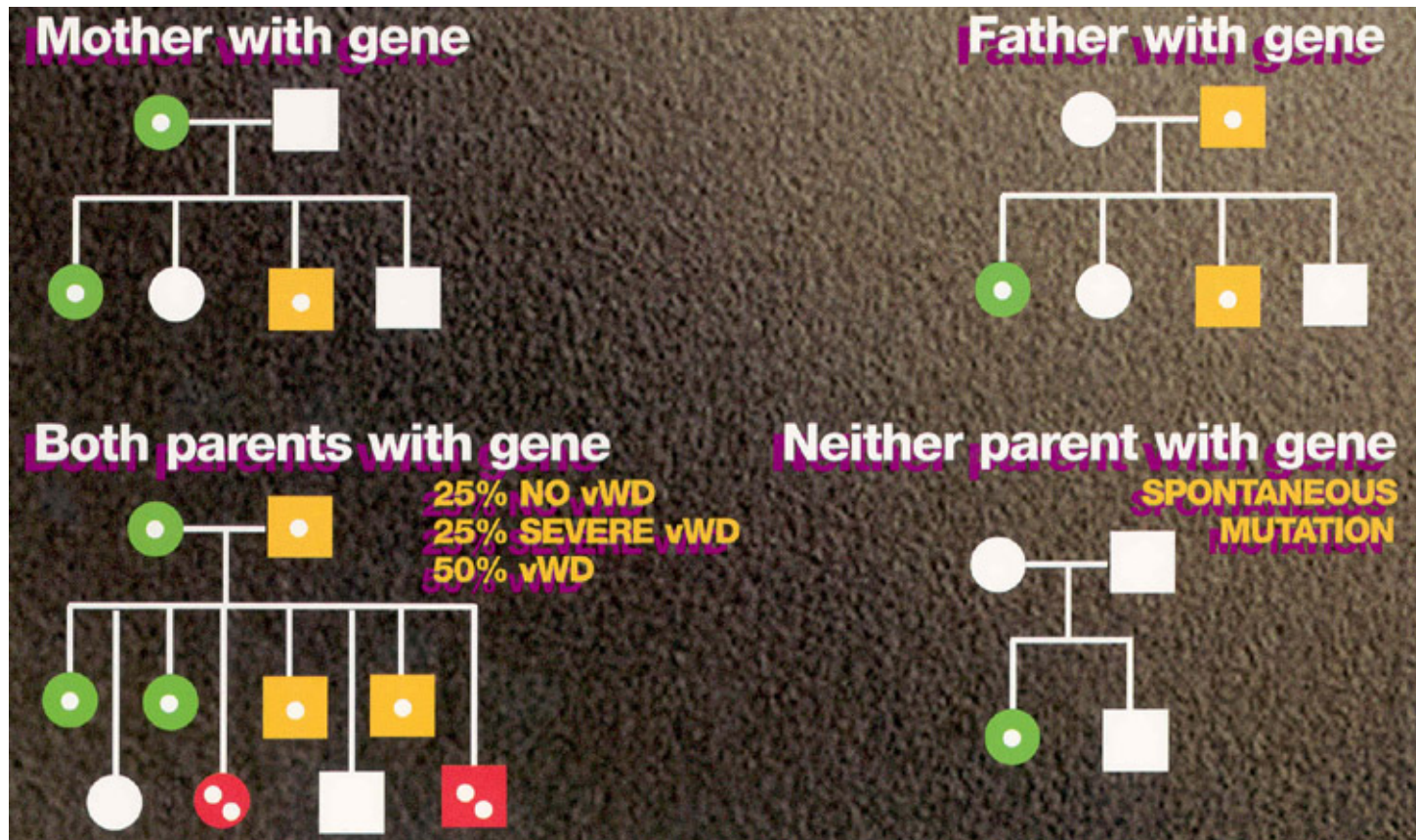


Inheritance

- The abnormal gene in vWD is on the autosomal chromosome, not on one of the sex-linked chromosomes as it is in hemophilia. Therefore, unlike hemophilia, which usually affects males, vWD affects both males and females in equal numbers.



Inheritance Diagram



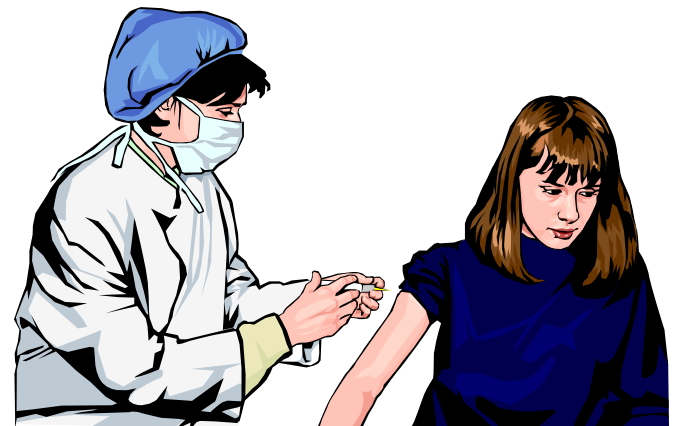
Signs and Symptoms

- Nose bleeds
- Mouth bleeding
- Gum bleeding
- Easy bruising
- Menorrhagia
- Mucous membrane bleeding
- Prolonged bleeding at the time of surgery
- Rare joint or muscle bleeding (except in severe type 3 vWD)



Diagnosis

- Diagnosis is made with a series of blood tests , usually ordered as a von Willebrand panel at a coagulation lab
- Diagnosis is made by an experienced hematologist

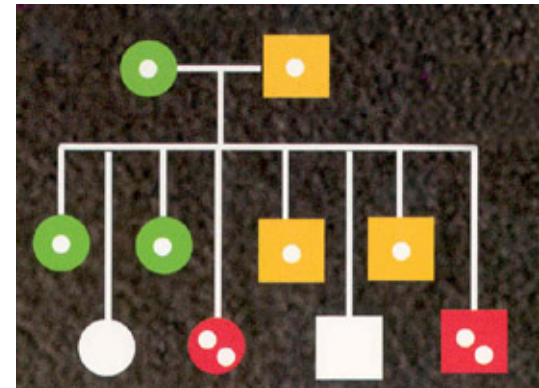


VWD Laboratory Testing

- Ristocetin Cofactor-evaluates how well VWF works
- Von Willebrand Factor Antigen-Measures amount of VWF present
- VWF Multimers-Measures size of VW protein. Helps classify type of VWD
- Factor VIII
- Platelet Function testing

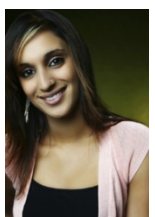
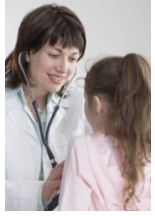
Diagnosis

- Since persons with vWD may occasionally have normal blood test results, family history becomes an important tool in the diagnosis



Types of von Willebrand Disease

- Type 1
- Type 2
- Type 3



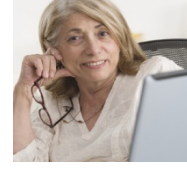
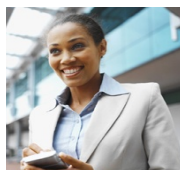
VWD Type 1

- Reduced levels of VWF
- The VWF that is present functions normally
- Mild and most common type



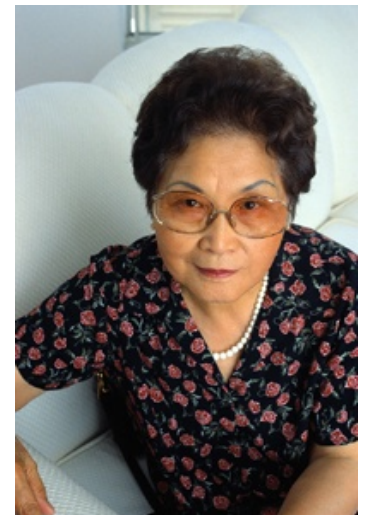
VWD Type 2

- There may be plenty of circulating VWF but it does not function properly
- May also have decreased levels of VWF, with normal or low factor VIII levels
- Subdivided into type 2a and 2b, type 2N (rare), or Platelet type



VWD Type 3

- Total or near absence of vWF with very low levels of factor VIII (3-10%)
- Causes frequent mouth, nose and GI bleeding
- Can cause severe joint and muscle bleeding, severe menorrhagia in women
- This is severe vWD



Question?

- Can a woman who is a carrier have bleeding symptoms?



Genetics

- Carrier women: can be symptomatic
- Carrier women: 50% chance with each pregnancy to have a son with hemophilia or daughter who is also a carrier
- May be spontaneous mutation



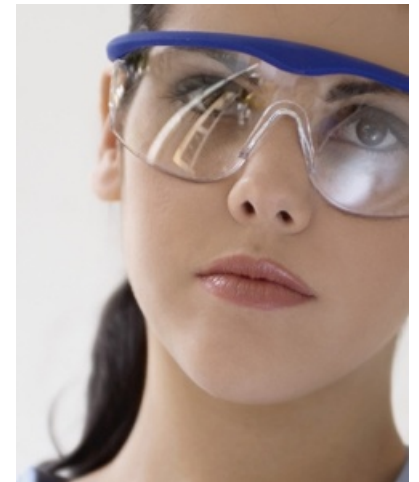
Mommy Facts

- DNA testing via amniocentesis between 9-15 weeks of gestation
- Known carrier with factor level $< 60\%$ should be tested in last trimester
- $< 60\%$ factor level should be corrected at term
- Delivery should be in a hospital



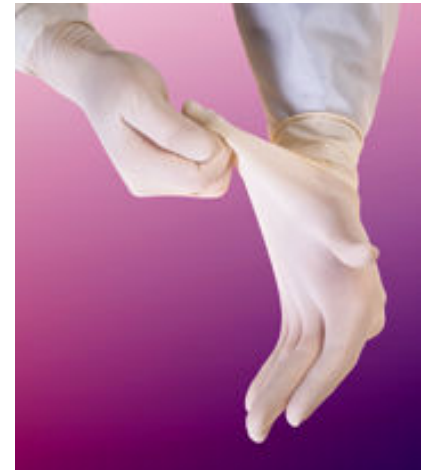
Universal Precautions

- To reduce the risk of transmission of HIV, Hepatitis and other blood borne infections
- Wear clean exam gloves when you may have direct contact with open skin, mucous membranes, blood and body fluids during any treatment and clean up

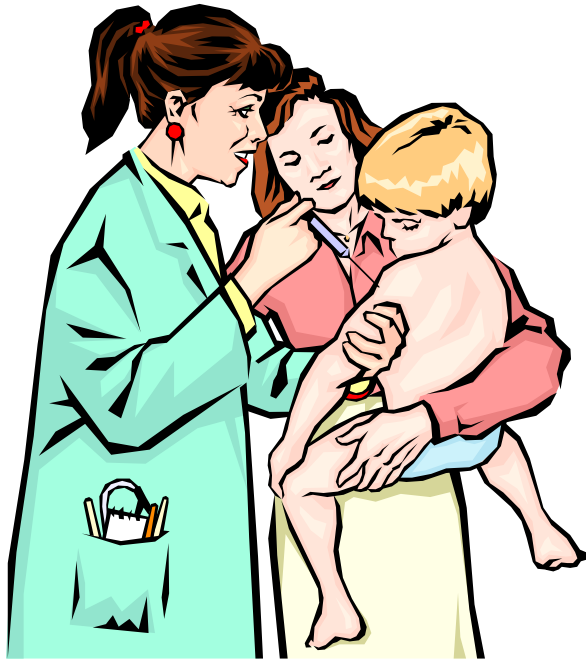


Universal Precautions

- Dispose of all supplies that are contaminated with blood and/or body fluids in the proper containers
- Wash your hands
- Wear clean disposable exam gloves



Treatment

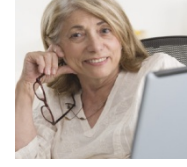


First Aid
Conservative treatment
Factor Replacement
Other medications



Bruising

- Treatment may not be necessary, depending on location of bruise and the type of VWD
- RICE (rest, ice, compression, elevation)



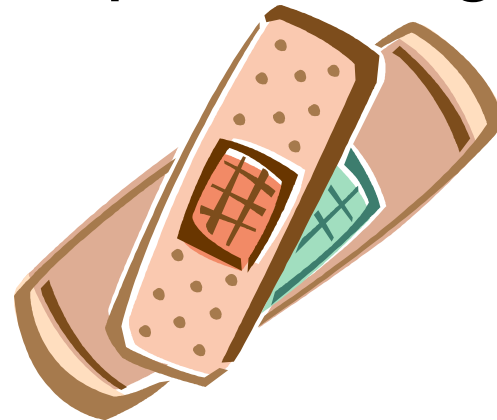
Nose Bleeds

- Put on gloves
- Pinch the nostrils for 10-15 minutes
- Ice pack to bridge of nose if necessary
- Keep head straight up



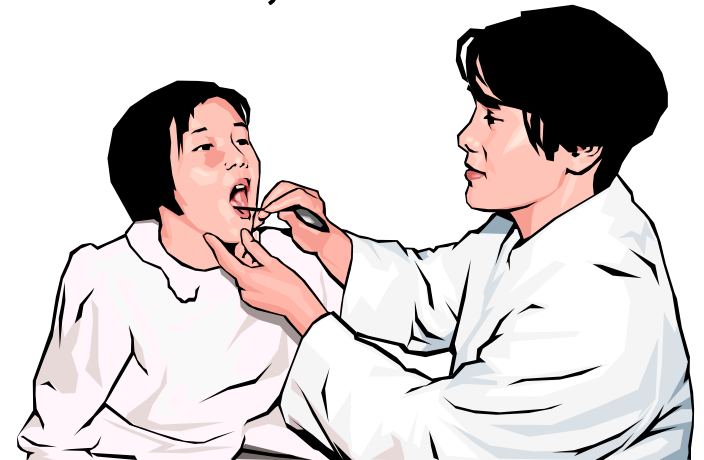
Abrasions

- Put on gloves
- Cleanse area with soap and water
- Apply pressure to stop bleeding, if necessary
- Apply bandage



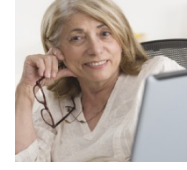
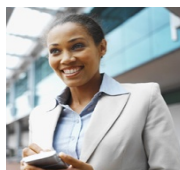
Mouth and Gum Bleeds

- Using gauze apply direct pressure
- Apply ice as needed
- Popsicle for young children
- Depending on type of VWD, may require other treatment



Menorrhagia

- Bed rest
- Birth control pills can be used to help control menorrhagia
- DDAVP nasal spray can also be used to control menses for some types of VWD
- Amicar is sometimes used in combination with other treatment



Life Threatening Bleeds

- Some types of severe bleeding can be life threatening
- Contact Hemophilia Treatment Center for assistance
- Physician's Hemophilia Hotline
 - 1-800-749-7468



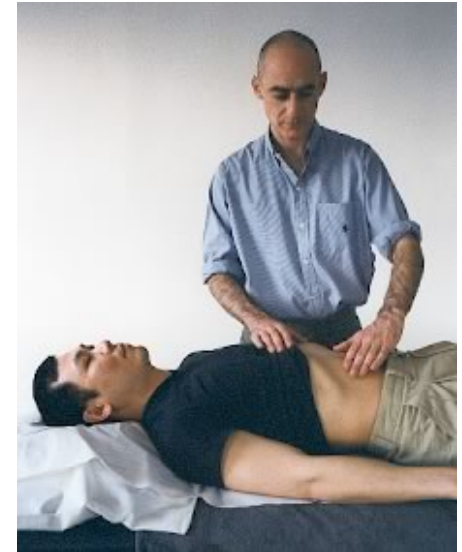
Life Threatening Bleeds

- Head injury – Monitor for loss of consciousness, nausea, vomiting, pain, blurred vision, sleepiness, signs of shock
- Throat – monitor for difficulty breathing or swallowing



Life Threatening Bleeds

- Abdominal injury – may be pale, with increased rigidity of abdomen, increased pulse, decreased blood pressure, symptoms of shock



Medication

- Blood products: Humate –P
 - rich in VWF and factor VIII
- Birth control (hormones)
- Lysteda
- Headache (not from an injury):
no aspirin, no NSAIDs



Medication

- Amicar: Antifibrinolytic agent- prevents the breakdown of a clot for soft tissue mouth and nose bleeds or preventive dental work.
- DDAVP: A synthetic hormone that can raise the levels of factor VIII for mild Hemophilia A or some types of von Willebrand Disease. Nasal spray (Stimate) or IV form available. Requires testing in non-bleeding state for response.

Amicar

Epsilon Amino Caproic Acid

- Antifibrinolytic
- Helps stabilize clots
- Dosing: 50 - 100 mg./kg. q. 6 hours
- Uses
 - Mucocutaneous bleeding

DDVAP

Desmopressin Acetate

- Synthetic vasopressin
- Administration
 - Intravenous
 - Subcutaneously
 - Nasally (Stimate)
- Method of action
 - release of stores from endothelial cells raising factor VIII and vWD serum levels
 - Side Effects

Medication

- DDAVP infusions for some types of VWD
 - must be tested in non-bleeding state
- DDAVP (Stimate) nasal spray
 - for some types of VWD
 - usually for minor bleeds



Factor Replacement

- 2 major types of factor
 - Plasma derived: heat and chemical treatment to prevent blood borne diseases
 - Recombinant: genetically engineered factor
- Treat within two hours of injury if possible
- Treat before procedures to prevent hemorrhage
- **WHEN IN DOUBT, TREAT!**
- On demand vs. Prophylaxis

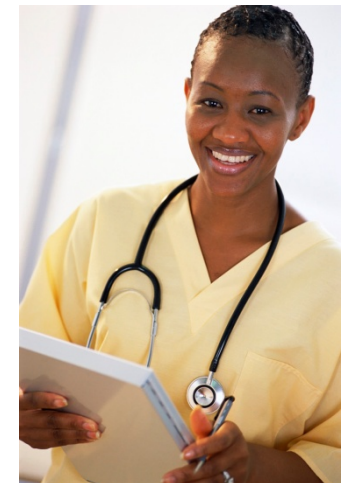
Factor Concentrates

- Factor VIII half life is 8 -12 hours and is gone by 48 hours
- Factor IX half life is 18 -24 hours and is gone by 72 hours
- All are given Intravenous - IV slow push
- Dose varies depending on type of bleed



Nursing Considerations

- Factor replacement to be given on time
- Laboratory monitoring
- Increase metabolic states will increase factor requirements
- Factor coverage for invasive procedures



Nursing Considerations

- Document - infusions, response to treatment
- Avoid NSAIDS, ASA
- Utilize Hemophilia Center staff for questions / problems
- Other agencies for support
- Genetic counseling and support



Psycho/Social Considerations

- Financial issues
- Insurance issues
- Guilt associated with Diagnosis
- Feeling different than others
- School/Work issues
- Control issues
- Confidentiality
- Other agencies referrals for support

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