

Bleeding Disorders for schools



LONE STAR
Bleeding Disorders Foundation

OBJECTIVES

- Introduction to Hemophilia and Von Willebrand disease
- Complications
- Discuss treatment options
- Introduce the Lone Star Chapter Bleeding Disorder Foundation



BLEEDING DISORDERS

Hemophilia A

Hemophilia B

Von Willebrand Disease



HEMOPHILIA

- Hemophilia A is a missing or reduced amount of Factor 8
- Hemophilia B is a missing or reduced amount of Factor 9
- The level of that factor in their blood determines severity
- X-linked inherited disorder, or a new mutation
- Up to 1,000 affected in the state of Texas
- Up to 33,000 living with hemophilia in the U.S.
- Hemophilia A is roughly 4 times more common than Hemophilia B



VON WILLEBRAND DISEASE

Missing/reduced or nonfunctioning Von Willebrand Factor

Males or females

Genetic inheritance

Most common bleeding disorder

Up to 1% of population or 1 in every 100

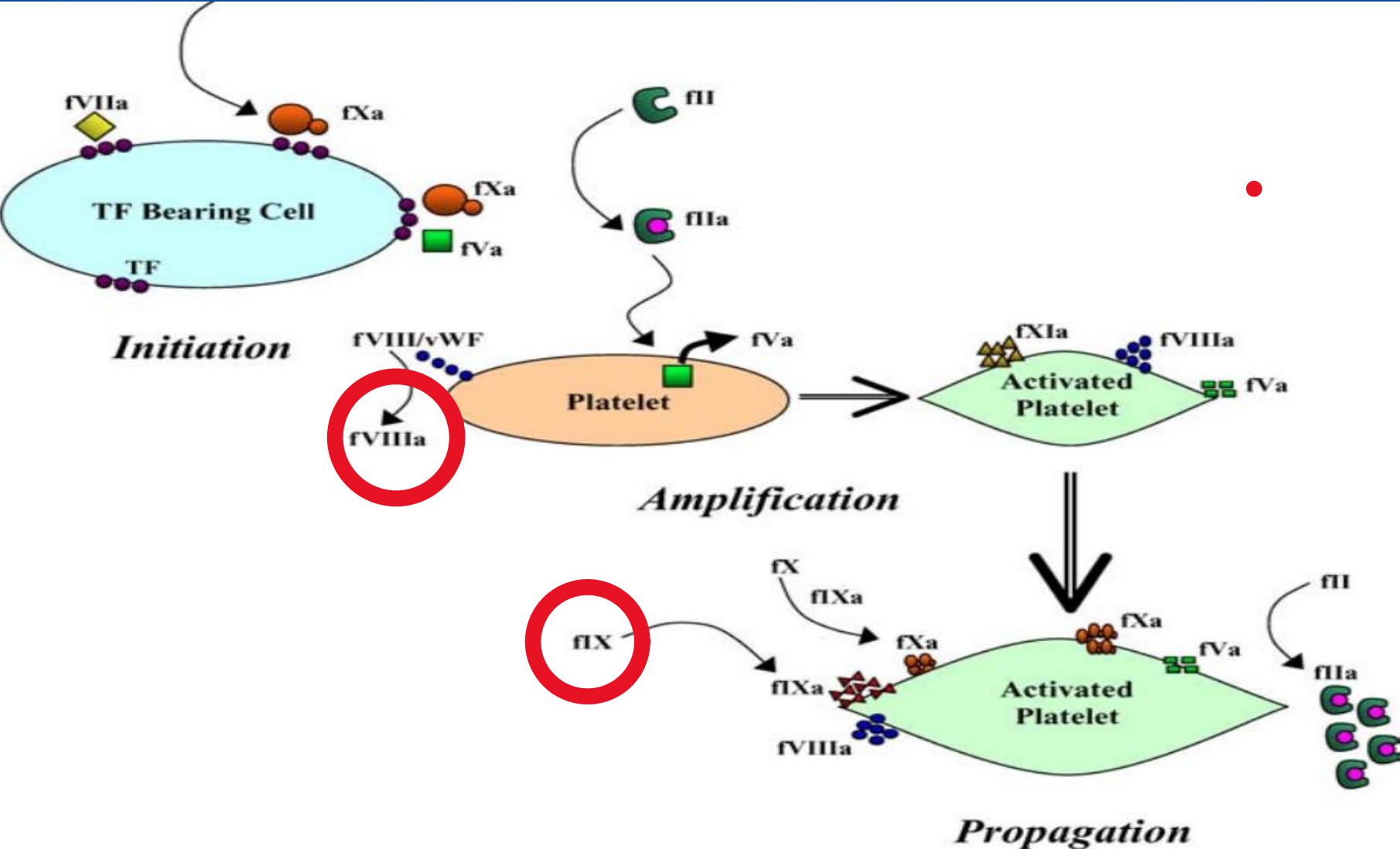
Many unaware they have it

Symptoms

- Nose and gum bleeding
- Menorrhagia (heavy menstrual bleeding)
- Bruising/hematomas
- Bleeding during procedures
- Uncommon, but can have joint and muscle bleeding

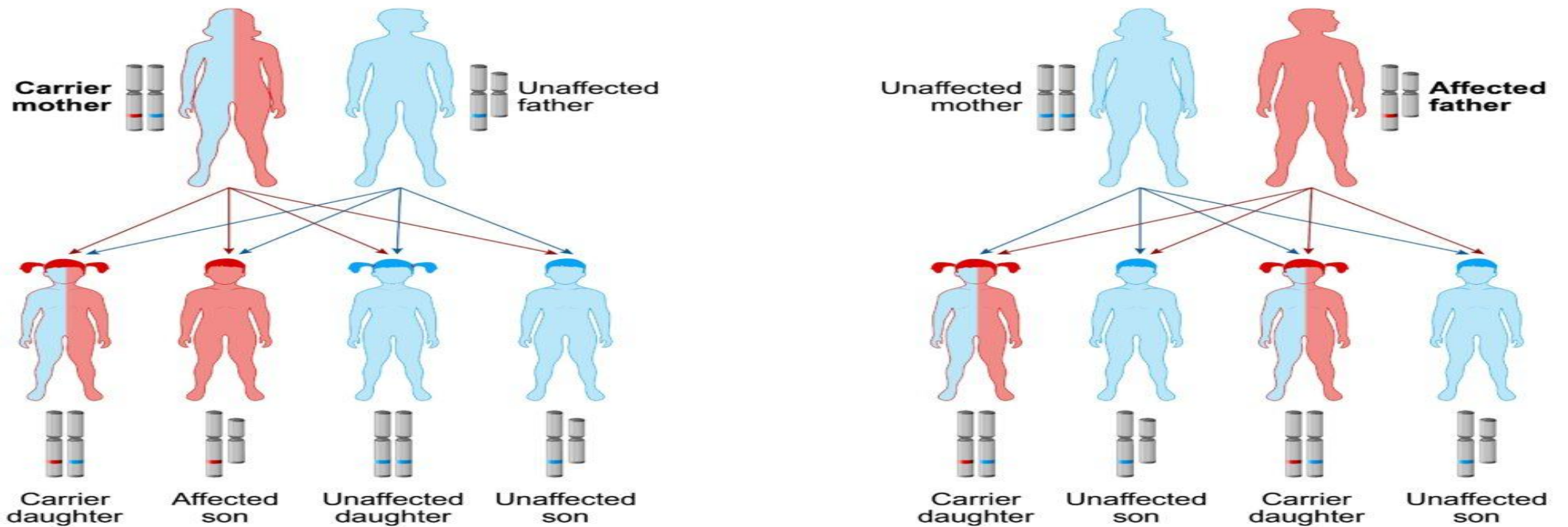


CLOTTING CASCADE



GENETICS OF HEMOPHILIA

X-linked recessive inheritance

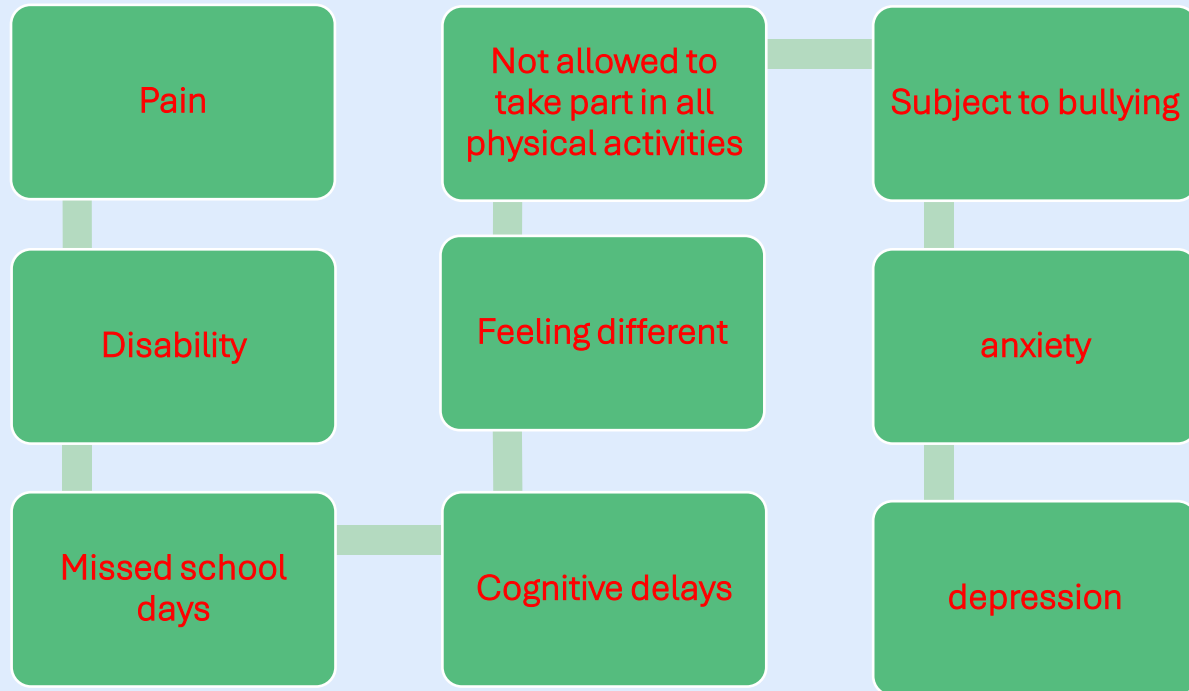


HEMOPHILIA A AND B

X-linked transmission:
mother passes gene
to her sons, carrier
status to her
daughters

Father passes carrier
status to his
daughters

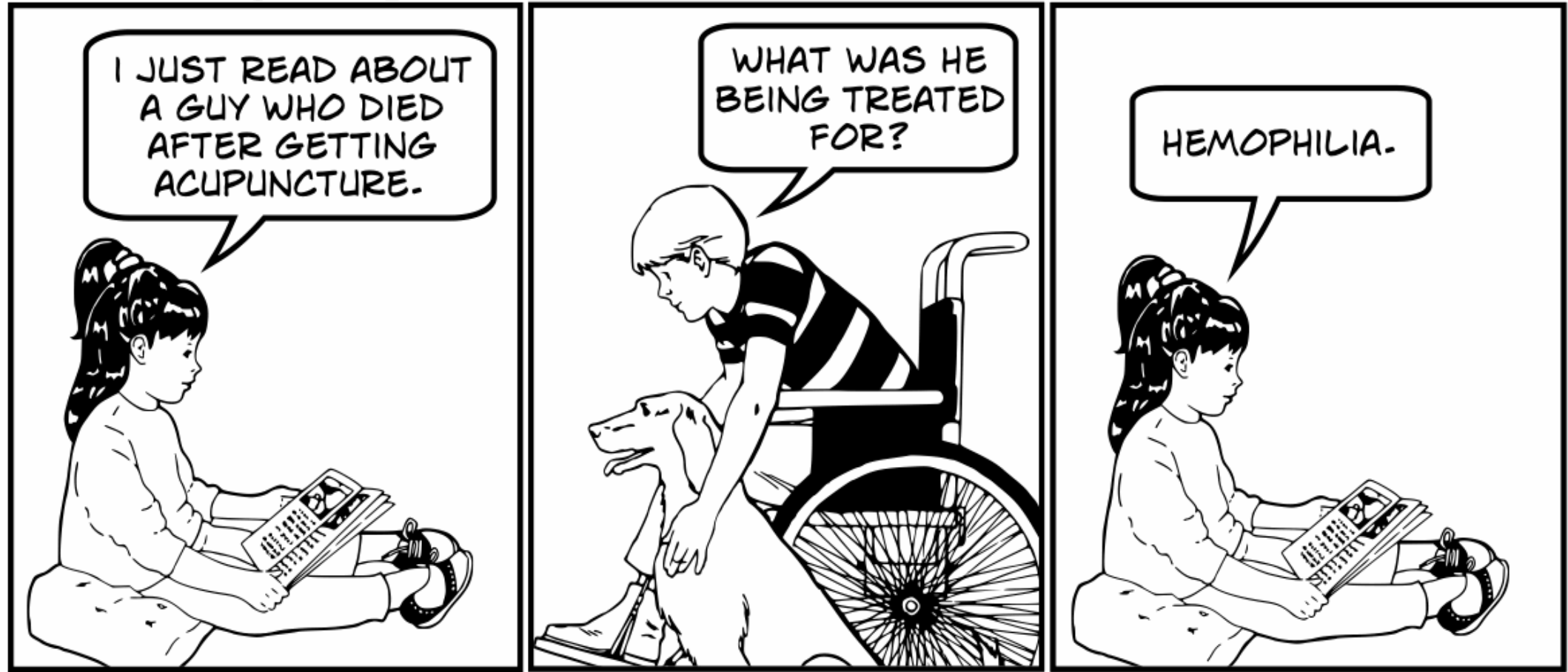
Carrier females can bleed
too



BLEEDING SYMPTOMS

Johnny Optimism

JohnnyOptimism.com / ©2015 by Stilton Jarlsberg



BLEEDING SYMPTOMS

- Bruising, hematomas
- Nose bleeding, gum bleeding
- Menorrhagia (heavy periods)
- Bleeding into muscles and joints
- Life threatening bleeding into head, neck, abdomen



BLEEDING SYMPTOMS

Can occur from:

- ~Injury/activity
- ~Invasive procedure/surgery
- ~Spontaneous or unexplained





BLEEDING INTO THE JOINT (HEMARTHROSIS)

- “Hallmark of hemophilia”
- Most common joints:
 - Knee
 - Ankle
 - Elbow
- Bleeding into the joint is:
 - Painful
 - Reduced range of motion
 - Inability to use/move the joint
 - Recurrent bleeding into a joint can result in a **lifelong disability**



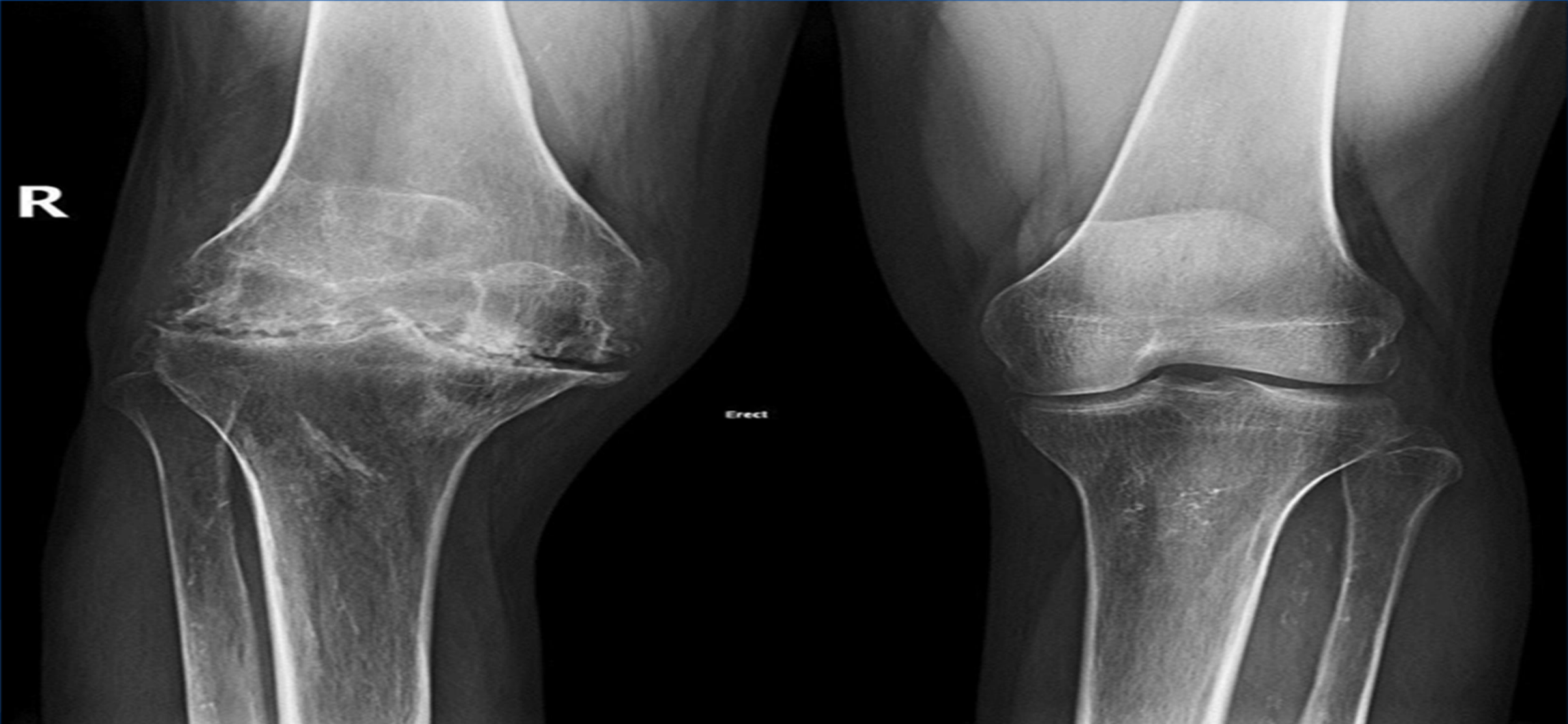


BLEEDING INTO THE JOINT (HEMARTHROSIS)

- Joint bleeds may **not** be visible to the eye
- Complaints of pain, feeling of joint “filling up”
- Will not want to use the affected extremity
- Treatment:
 - **Factor** replacement therapy
 - **RICE** (rest, ice, compression and elevation)



HEMOPHILIA ARTHROPATHY





BLEEDING INTO THE MUSCLE

- Any muscle can experience bleeding
 - Pain
 - Disability
 - Decreased ROM
 - With or without swelling or bruising
 - Forearm and calf bleed can result in **compartment syndrome**
 - iliopsoas muscle in the pelvis can cause extreme **pain, bent over walking**, blood loss leading to **anemia**
- Treatment: Factor replacement therapy and
- RICE

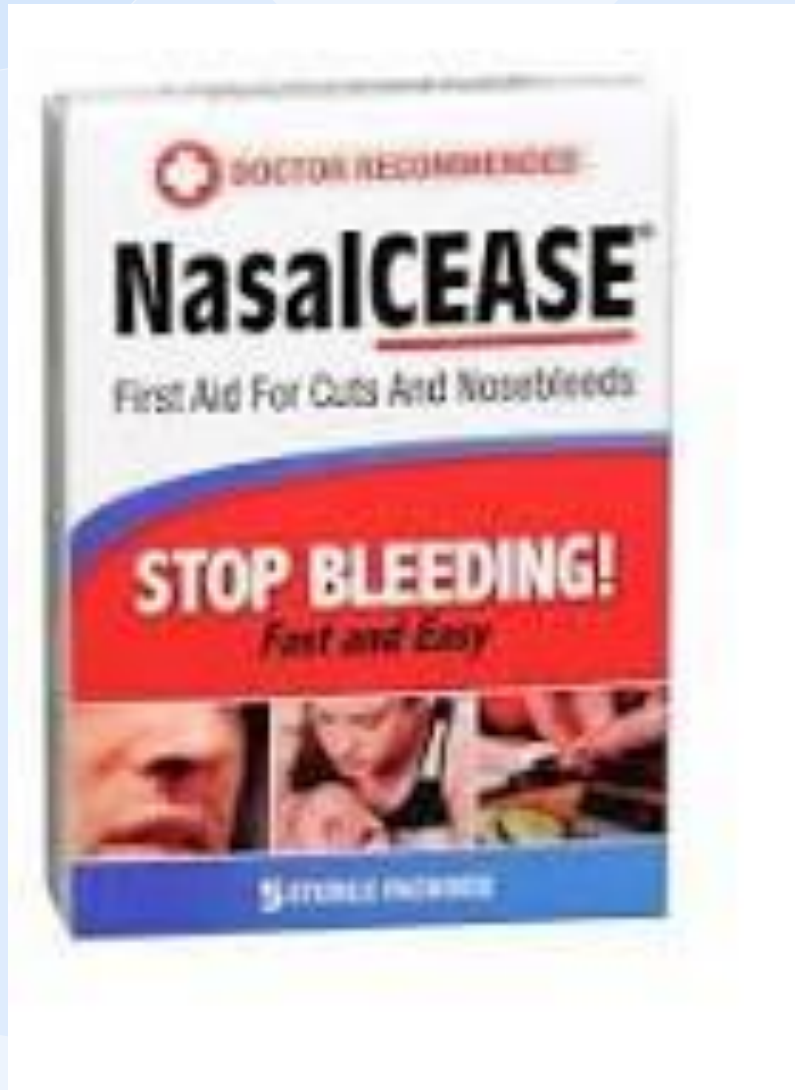




NOSEBLEEDS (EPISTAXIS)

- Chronic
- Recurring
- Can result in anemia, sometimes severe
- Upset stomach/vomiting from swallowing blood





NOSEBLEEDS (EPISTAXIS)

- How to stop:
 - Pressure and/or ice to bridge of nose
 - Lean forward
 - Nose packing with gauze or NasalCease
- Medications such as:
 - Aminocaproic Acid
 - Tranexamic acid
 - Afrin nasal spray
 - Antihistamines
 - Nasal saline
 - Vaseline



GINGIVAL BLEEDING

Can occur:

- After tooth loss
- Dental procedures
- Trauma

- Treatment:
 - Pressure
 - Ice
 - Cold beverages, ice cream, popsicles
 - May require dental intervention
 - Aminocaproic or tranexamic acid



MENORRHAGIA

Heavy menstrual bleeding:

- Lasting >8 days
- Consistently soaks through 1 or more sanitary protections every 2 hours on multiple days
- Requires use of >1 sanitary protection item at a time
- Requires changing sanitary protection during the night
- Associated with repeat passing of blood clots



MENORRHAGIA

Things to consider:

- Girls/women are often not recognized as having a bleeding disorder
- Heavy menstrual bleeding may be seen as normal in her family
- Girls in school may need access to the restroom more frequently due to frequent need to change pads/tampons
- Leakage onto clothes may be frequent
- May need to miss school due to amount of bleeding
- Can cause anemia, sometimes severe



LIFE THREATENING BLEEDS

- **Neck/throat** bleeds/injuries: can be life threatening as swelling/bleeding can occlude the airway
- **Intracranial/intraspinal:** life threatening injuries requiring factor replacement immediately.
- **Abdominal:** Injury to abd or severe abdominal pain causing vomiting of blood, dark tarry stools, bright red stools, abd distension. Bleeding can be excessive and life threatening
- **Eye injuries:** can cause loss of vision
- **Kidney:** painless blood in urine of unknown cause, passing blood clots can cause pain, blood clot occlusion can cause kidney injury
- **Notify parent first, hematologist and decide if EMS is required. Student will need to be given their factor replacement product**



TREATMENT

- Typically requires **very expensive IV replacement** factor infusions over the entire lifespan
- Could also be on a subcutaneous injection to prevent bleeding
- Individualized, dynamic and very specialized
- Patients/families are taught how to manage their disease at home by learning to infuse their factor.
- Poor understanding of bleeding disorders in the medical community
- Patients usually go to a federally funded hemophilia treatment center (San Antonio, Austin, Houston or Dallas)



MEDICATIONS FOR PREVENTION AND TREATMENT OF BLEEDING

- Subcutaneous and intravenous products



FACTOR INFUSION

I don't think there's a
'typical' person with hemophilia.

- Cathy
hemophilia mom



ACTIVITIES

- Usually discourage contact sports but
- Some may play baseball, basketball, soccer
- Football, wrestling, bull riding, hockey is usually a NO due to risk of head injuries
- Dodge ball, high jungle gyms, trampoline and trampoline parks are also usually discouraged
- Can be active in many sports such as tennis, golf, swimming, track
- Should be active and included as much as possible
- Protective gear is a must
- Parents should know how to manage bleeding symptoms when and to call the hematologist



LONE STAR BLEEDING DISORDER FOUNDATION

- 501 (c) (3) non-profit organization to provide support to persons and families with a bleeding disorder in South Texas
- Started in 1969, incorporated in 1991
- Supports over 700 families

ACTIVITIES

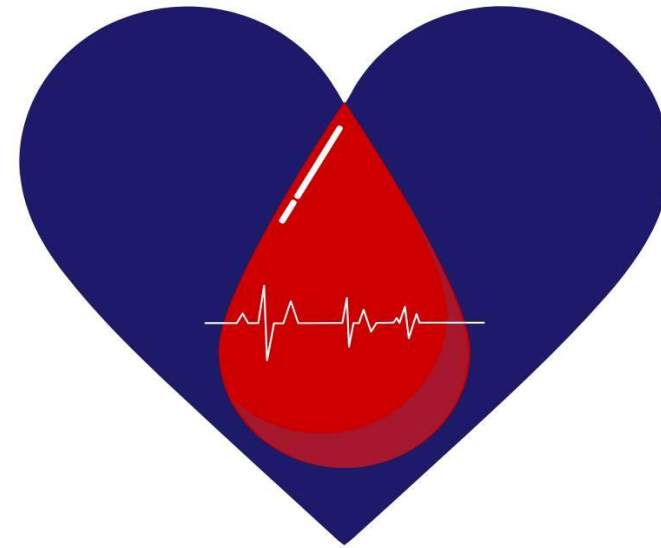
- Education: Annual Texas Bleeding Disorder Conference and other smaller events
- Advocacy: Washington Days and Capital Days
- Development of community
- Improve access to care
- Fundraising :helps support the Texas Hemophilia week- long summer camp
- Helping Hands : emergency financial assistance
- Website: Lonestarbleedingdisorders.org



Thank you!

Trish Amerson

Outreach Coordinator
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NATIONAL
BLEEDING DISORDER
AWARENESS MONTH