Emergencies in Hemophiliacs

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ABSTRACT:

Audience: This modified team-based learning (mTBL) session is appropriate for medical students or emergency medicine residents.

Introduction: Hemophilia is an x-linked recessive disorder leading to a decrease in functional clotting factors; there are two types, Hemophilia A and B, with deficiencies in clotting factors VIII and IX respectively. Patients are typically male and suffer from recurrent episodes of bleeding. The prevalence of hemophilia is low; therefore, many residents may not treat a patient with hemophilia during their training. Understanding the management and dosing of treatment factors is essential for emergency physicians.

Objectives: By the end of this session learners will be able to: 1) describe the underlying deficiencies of hemophilia A and B; 2) discuss the complications of hemophilia; 3) formulate an appropriate treatment plan for an acutely bleeding hemophiliac; 4) calculate the appropriate factor dosing for a hemophiliac with acute bleeding.

Method: The format of this education session is a modified TBL.

Topics: Hematology, hemophilia, bleeding emergencies, hemorrhage.
Linked objectives, methods and results:
The two Learner Responsible Content articles provide an overview of the background, complications, and treatment for hemophilia, after which learners will be able to describe the underlying deficiencies in hemophilia A and B and discuss the complications and treatment options for hemophilia.

The modified TBL (mTBL) consists of 2 portions, a learning worksheet (Hemophilia Worksheet, Parts A-C) and the “Group Application Exercises.”

The “Hemophilia Worksheet” Parts A-C will take approximately 30 minutes for learners to complete. This can be done individually or in small groups. The worksheet will test the learners’ understanding of hemophilia and its complications.

Learners will also formulate a treatment plan and calculate factor dosages for acute treatment for a bleeding hemophiliac. The Group Application Exercises consist of four cases that focus on emergent treatment in a bleeding hemophiliac. Groups will need to choose the correct factor and calculate dosages for each case. This portion should take approximately 15 minutes.

Recommended pre-reading for instructor:

Learner responsible content (LRC):
Any of the following:

Results and tips for successful implementation:
• If your learners do not have access to the content prior to the session, they can access the two free open-access medical education sites listed above during the group activity.
• Instructors should assign groups; learners should not self-select. The session is most successful with groups of four to five learners.

Implementation:
1. Three to seven days before the didactic session, instruct learners to read one or more of the above suggested
readings. The instructor should also read one or more of the suggested readings.

2. If using the “Hemophilia Worksheet” Parts A-C as an individual readiness assessment test (iRAT), print one copy of the “Hemophilia Worksheet” for each individual. If using the worksheet as a group readiness assessment test (gRAT), print one copy per group. If using it for both iRAT and gRAT, print 1 copy per learner and 1 copy per group.

3. Print one copy of the “Group Application Exercises” for each group.

4. Print one copy of the “Hemophilia Worksheet Parts A-C Answers”, and “GAE Answers” for each instructor.

Content:
- iRAT and gRAT: Hemophilia Worksheet Parts A-C
- GAE
- RAT Key: Hemophilia Worksheet Parts A-C
- GAE Key

References/suggestions for further reading:


Individual Readiness Assessment Test (iRAT) and Group Readiness Assessment Test (gRAT)

The iRAT and gRAT (Hemophilia Worksheets Part A, B and C) start on the following page and be completed individually or as a group.
## Hemophilia Worksheet, Part A

<table>
<thead>
<tr>
<th></th>
<th>Hemophilia A</th>
<th>Hemophilia B</th>
<th>von Willebrands</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>How is it inherited</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Most Commonly Affects: Males, Females or Both?</strong></td>
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<tr>
<td><strong>Factor Deficiency</strong></td>
<td></td>
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<tr>
<td><strong>Prevalence</strong></td>
<td></td>
<td></td>
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<tr>
<td><strong>% of Hemophilia Cases</strong></td>
<td></td>
<td></td>
<td>n/a</td>
</tr>
<tr>
<td><strong>Prolonged PT, PTT or Bleeding time?</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Treatment?</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Hemophilia Worksheet, Part B: FACTOR LEVELS & BLEEDING

Normal plasma levels of factor VIII or IX range from _____ to _____%.

Persons with < _____% factor VIII or IX activity are considered to have SEVERE hemophilia. Frequent bleeding episodes are (pick one) common / uncommon. Bleeding can occur (pick one) spontaneously / after minor trauma / only after significant trauma or surgery. The most common location of bleeding is .

Persons with _____% factor VIII or IX activity are considered to have MODERATE hemophilia. Persons may experience bleeding (pick one) spontaneously / after minor trauma / only after significant trauma or surgery, but should not bleed (pick one) spontaneously / after minor trauma / only after significant trauma or surgery.

Persons with more than ______% factor VIII or IX activity are considered to have MILD hemophilia. These patients typically bleed (pick one) spontaneously / after minor trauma / only after significant trauma or surgery.

The FIVE major sites of serious bleeding.
(think life, limb, function threatening)
1. 
2. 
3. 
4. 
5. 

The joints most commonly affected by bleeding:
1. 
2. 
3. 

**THESE ALL REQUIRE IMMEDIATE ASSESSMENT AND TREATMENT AS THEY ARE CHARACTERIZED BY BLEEDING INTO AN ENCLOSED SPACE, COMPRESSION OF VITAL TISSUE, POTENTIAL LOSS OF LIFE LIMB OR FUNCTION.**

What is the most common cause of death in patients with hemophilia?

Of note, repeated hemorrhage is a risk factor for future hemorrhage, as this leads to thickened synovial tissue and friable blood vessels.

**Hemophilia Worksheet, Part C: FACTOR TREATMENT OPTIONS**

<table>
<thead>
<tr>
<th>Severity of Bleed</th>
<th>Example of Bleed (e.g. location or bleeding or type of trauma)</th>
<th>Goal Factor Activity %</th>
<th>Repeat Dosing?</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moderate</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Severe</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**FACTOR TYPES:**

Match the 3 types of factor replacements to the correct definition.

Options: *Intermediate Purity, Recombinant, Purified*

- ___________: Non-human product
- ___________: Human product, only contains factor VIII
- ___________: Human product, contains factor VIII, also contains vWF

**1° PPX vs 2° PPX**

- **Primary Prophylaxis:** Scheduled factor replacement dosing, typically done at home or outpatient for severe hemophiliacs to reduce the likelihood of spontaneous bleeding.
- **Secondary Prophylaxis:** Emergent or scheduled dosing of factor replacement in response to an injury or in anticipation of a surgery, ie replacement for an acute need.
HEMOPHILIA A Treatment Options

Factor ____
Treatment of choice for moderate to severe disease in Hemophilia ____
Factor VIII = Each U/kg = an Increase of Factor activity by ____
____ U/kg = an Increase of Factor activity to 100%

DDAVP
Use in patients that have known Factor Activity >10%, who have mild bleeds
Use in patients who can tell you it has worked for them before
DOSE: ____ mcg/kg/dose IV

Cryoprecipitate
Contains Factor VIII and vWF
Use only if no other options available
DOSE: 1 bag per ____ kg of body weight

TXA or Amiocaproic Acid
Helps stabilize clot formation
Only for mucosal bleeds
NEVER in patients with renal bleeds or renal insufficiency.
TXA DOSE: ____ mg/kg IV or____ mg/kg PO q 6-8 hrs, can be used topically as well

HEMOPHILIA B Treatment Options

Factor ____
Treatment of choice for moderate to severe disease in Hemophilia ____
Factor IX = Each U/kg = an Increase of Factor activity by ____
____ U/kg = an Increase in Factor activity to 80%

TXA or Amiocaproic Acid
Helps stabilize clot formation
Only for mucosal bleeds
NEVER in patients with renal bleeds or renal insufficiency.
TXA DOSE: ____ mg/kg IV or____ mg/kg PO q 6-8 hrs, can be used topically as well

Fresh Frozen Plasma
Contains Factor IX
Only use if you have NO other options, as can lead to ________________________
DOSE: ____ mL/kg initially
TREAT

Instructions: Fill in the _____, then use this sheet to help answer the group application questions below.

1. Factor Level:
   a. Option A: The patient knows their factor activity level
   b. Option B: Patient Does Not Know their factor level→Assume a factor level of _____
   c. Option C: In the ED always assume factor level of 0%

2. Consider the degree of bleed, how much activity do you need to get them to?

3. Patient’s Weight

4. Which factor are you giving? Hemophilia A or B?

5. Calculate out how much to give the patient.
   Desired Factor Level% – Patients Factor Activity% = How much Factor they need%

   Factor VIII = Each U/kg = an Increase of Factor activity by _____
   ______ U/kg = 50% correction (for minor bleeds)
   ______ U/kg = 100% correction (for major bleeds)

   Factor IX = Each U/kg = an Increase of Factor activity by _____
   ______ U/kg = 50% correction (for minor bleeds)
   ______ U/kg = 100% correction (for severe bleeds)

   FACTOR NEEDED = Wt in kg x How much Factor they need% x U/kg to get to 100%

   MATH: Patient has a “severe bleed”, they do not know their activity level.
   Goal 100% - 0% Unknown = Need 100% Factor Activity Level Change
   Factor VIII = Each U/kg = an Increase of Factor activity by _____
   Factor IX = Each U/kg = an Increase of Factor activity by _____

   For a 50 kg patient:
   Hemophilia A: 50kg x _____U/kg = _____U needed to get to 100%
   Hemophilia B: 50kg x _____U/kg = _____U needed to get to 100%

6. DO NOT WAIT TO HAVE A DIAGNOSIS, IF THE PATIENT THINKS THEY HAVE A BLEED, DOSE. GET CONFIRMATORY TESTS LATER AND CALL HEMATOLOGY AFTER 1ST DOSE IS GIVEN.

7. If the family bring their factor with them use it! It will save time, and for patients with inhibitors the factor they bring may be the only one that works for them!

8. Do not waste factor, it is $$$$, use the closest vial size to the recommended dose, and give the whole vial. Giving too much factor will ensure it lasts longer.

9. Ask the patient or parents questions, they will likely know more than you about their illness and can answer questions.

10. Factor needs to be given in its own line, it can not be given with anything else! Typically, you will use a butterfly needle to give it.
A 13 yo M presents with bleeding gums, he states that he has intermittent nose bleeds, and that he believes his family has an unknown bleeding disorder. Which of the following disorders is most likely?

35 y/o M with hemophilia A presents with right knee pain. He says that he believes he is bleeding in his knee. He weighs 80 kg, and believes his factor level is <2%. How much factor must be used to reach 40% activity?

A 12 y/o M with hemophilia B presents with complaints of getting hit in the flank by a basketball just prior to arrival. He states that he has severe pain on that side. He doesn’t know his factor level. He weighs 35kg. How much factor should the patient receive?

A 22 y/o M with Hemophilia A presents with complaints of gum bleeding x 2 hours. He states that he was unable to give himself factor at home but is concerned. He states that he has a factor level of 5% and requests that you raise his level to 50%. He states that he weighs 75kg.
BONUS: ACQUIRED HEMOPHILIA Information

These are patients with inhibitors to clotting factors making treatment more difficult.

Usually Caused by:
1. Malignancies (CLL, Adenocarcinomas)
2. Pregnancy or Postpartum State
3. Autoimmune Disorders (SLE, RA)
4. Idiopathic

If you are unsure if the patient has Factor Inhibitors, you can perform the Clotting Factor Mixing Test
- Mix normal plasma with the patient’s serum
- If the patient is Normal and does not have Acquired Hemophilia= aPTT will normalize
- If the patient has Acquired Hemophilia or Factor Inhibitors = aPTT remains prolonged

Low titers of inhibitor: treat with Factor VIII concentrates

High titers of inhibitor: treat with factor VIIa concentrates or prothrombin complexes (PCCs)

TREATMENT FOR PATIENTS WITH INHIBITORS:

Recombinant Factor VIIa: “bypasses” the inhibitors.
- Dose: 90-150 mcg/kg
- Has a short half-life and may require multiple additional doses

Factor Eight Inhibitor Bypassing Agent: is a plasma-derived bypassing agent.
- Dose: 50 U/kg
  Caution: Must be given slowly, because rapid infusion can cause excessive clotting (e.g. stroke, limb ischemia, etc).


INSTRUCTOR MATERIALS

Answer keys to all exercises with explanations, are on the following pages.

Learners: please do not proceed.
### Hemophilia Worksheet, Part A Key

<table>
<thead>
<tr>
<th>How is it inherited</th>
<th>Hemophilia A</th>
<th>Hemophilia B</th>
<th>von Willebrands</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>x-linked recessive</td>
<td>x-linked recessive</td>
<td>Type I and II AD Type III AR</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Most Commonly Affects: Males, Females or both?</th>
<th>Males</th>
<th>Males</th>
<th>Both</th>
</tr>
</thead>
<tbody>
<tr>
<td>Females affected if mom is carrier and dad has disease or if XO (Turners)</td>
<td>Males affected if mom is carrier and dad has disease or if XO (Turners)</td>
<td>Both</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Factor Deficiency</th>
<th>Factor VIII deficiency</th>
<th>Factor IX deficiency</th>
<th>vWF required for plt adhesion</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Prevalence</th>
<th>1/5000</th>
<th>1/30,000</th>
<th>1/100 asymptomatic 1/10,000 symptomatic</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>% of Hemophilia Cases</th>
<th>80%</th>
<th>20%</th>
<th>n/a</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Prolonged PT, PTT or Bleeding time?</th>
<th>PTT</th>
<th>PTT</th>
<th>PTT Bleeding Time</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Treatment?</th>
<th>Factor VIII</th>
<th>Factor IX</th>
<th>DDAVP, aminocaproic acid, tranexamic acid, and factor replacement</th>
</tr>
</thead>
</table>
Hemophilia Worksheet, Part B Key:

**FACTOR LEVELS & BLEEDING**

Normal plasma levels of factor VIII or IX range from **50% to 150%**.

Persons with **<1%** factor VIII or IX activity are considered to have SEVERE hemophilia. Frequent bleeding episodes are (pick one) **common**. Bleeding can occur (pick one) **spontaneously**. The most common location of bleeding is **into the joints**.

Persons with **1-5%** factor VIII or IX activity are considered to have MODERATE hemophilia. Persons may experience bleeding (pick one) **after minor trauma**, but should not bleed (pick one) **spontaneously**.

Persons with more than **5%** factor VIII or IX activity are considered to have MILD hemophilia. These patients typically bleed (pick one) **only after significant trauma or surgery**.

The FIVE major sites of serious bleeding.
(think life, limb, function threatening)

6. **INTRACRANIAL/SPINAL CORD**
7. **AIRWAY**
8. **GI/INTRA-ABDOMINAL**
9. **LIMB COMPARTMENTS**
10. **OCULAR**

The most frequent joint bleeds are:

4. **ELBOWS**
5. **KNEES**
6. **ANKLES**

**THESE ALL REQUIRE IMMEDIATE ASSESSMENT AND TREATMENT AS THEY ARE CHARACTERIZED BY BLEEDING INTO AN ENCLOSED SPACE, COMPRESSION OF VITAL TISSUE, POTENTIAL LOSS OF LIFE LIMB OR FUNCTION.**

What is the most common cause of death in patients with hemophilia?

**INTRACRANIAL HEMORRHAGE**

*Of note, repeated hemarthrosis is a risk factor for future hemarthrosis, as this leads to thickened synovial tissue and friable blood vessels.*
**Hemophilia Worksheet, Part C Key:**

**FACTOR TREATMENT OPTIONS**

<table>
<thead>
<tr>
<th>Severity of Bleed</th>
<th>Example of Bleed (e.g. location or bleeding or type of trauma)</th>
<th>Goal Factor Activity %</th>
<th>Repeat Dosing?</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>Minor trauma, hemarthrosis</td>
<td>30-40%</td>
<td>Single dose ok!</td>
</tr>
<tr>
<td>Moderate</td>
<td>Oral or mucosal bleeding Epistaxis Prophylaxis before surgical/dental procedure</td>
<td>50-80%</td>
<td>Consider observation and repeat dosing to activity 50% q12 x 1</td>
</tr>
<tr>
<td>Severe</td>
<td>Severe Trauma, Head, Neck or Abdomen trauma</td>
<td>80-100%</td>
<td>Repeat to 50% activity q12</td>
</tr>
</tbody>
</table>

**FACTOR TYPES:**

Match the 3 types of factor replacements to the correct definition.

- **Recombinant:** Non-human product
- **Purified:** Human product, only contains factor VIII
- **Intermediate purity:** Human product, contains factor VIII, also contains vWF

**1° PPX vs 2° PPX**

- **Primary Prophylaxis:** Scheduled factor replacement dosing, typically done at home or outpatient for severe hemophiliacs to reduce the likelihood of spontaneous bleeding.
- **Secondary Prophylaxis:** Emergent or scheduled dosing of factor replacement in response to an injury or in anticipation of a surgery, i.e., replacement for an acute need.
HEMOPHILIA A Treatment Options

Factor VIII
Treatment of choice for moderate to severe disease in Hemophilia A
Factor VIII = Each U/kg = an Increase of Factor activity by 2%
50 U/kg = an Increase of Factor activity to 100%

DDAVP
Use in patients that have known Factor Activity >10%, who have mild bleeds
Use in patients who can tell you it has worked for them before
DOSE: 0.3 mcg/kg/dose IV

Cryoprecipitate
Contains Factor VIII and vWF
Use only if no other options available
DOSE: 1 bag per 6 kg of body weight

TXA or Amicaproic Acid
Helps stabilize clot formation
Only for mucosal bleeds
NEVER in patients with renal bleeds or renal insufficiency.
TXA DOSE: 10 mg/kg IV or 25 mg/kg PO q 6-8 hrs, can be used topically as well

HEMOPHILIA B Treatment Options

Factor IX
Treatment of choice for moderate to severe disease in Hemophilia B
Factor IX = Each U/kg = an Increase of Factor activity by 1%
80 U/kg = an Increase in Factor activity to 80%

TXA or Amicaproic Acid
Helps stabilize clot formation
Only for mucosal bleeds
NEVER in patients with renal bleeds or renal insufficiency.
TXA DOSE: 10 mg/kg IV or 25 mg/kg PO q 6-8 hrs, can be used topically as well

Fresh Frozen Plasma
Contains Factor IX
Only use if you have NO other options, as can lead to volume overload
DOSE: 15 mL/kg initially
INSTRUCTOR MATERIALS

TREAT

Instructions: Fill in the _____, then use this sheet to help answer the group application questions below.

11. Factor Level:
   a. Option A: The patient knows their factor activity level
   b. Option B: Patient Does Not Know their factor level
      ➔ Assume a factor level of 0%
   c. Option C: In the ED always assume factor level of 0%

12. Consider the degree of bleed, how much activity do you need to get them to?

13. Patient’s Weight

14. Which factor are you giving? Hemophilia A or B?

15. Calculate out how much to give the patient.
   Desired Factor Level% – Patients Factor Activity% = How much Factor they need%

   Factor VIII = Each U/kg = an Increase of Factor activity by 2%
   25 U/kg = 50% correction (for minor bleeds)
   50 U/kg = 100% correction (for major bleeds)

   Factor IX = Each U/kg = an Increase of Factor activity by 1%
   50 U/kg = 50% correction (for minor bleeds)
   100 U/kg = 100% correction (for severe bleeds)

   **FACTOR NEEDED = Wt in kg x How much Factor they need% x U/kg to get to 100%**

   MATH: Patient has a “severe bleed”, they do not know their activity level.
   Goal 100% - 0% Unknown = Need 100% Factor Activity Level Change
   Factor VIII = Each U/kg = an Increase of Factor activity by 2%
   Factor IX = Each U/kg = an Increase of Factor activity by 1%

   For a 50 kg patient:
   * Hemophilia A: 50kg x 50U/kg = 2500U needed to get to 100%
   * Hemophilia B: 50kg x 100U/kg = 5000U needed to get to 100%

16. DO NOT WAIT TO HAVE A DIAGNOSIS, IF THE PATIENT THINKS THEY HAVE A BLEED, DOSE. GET CONFIRMATORY TESTS LATER AND CALL HEMATOLOGY AFTER 1ST DOSE IS GIVEN.

17. If the family bring their factor with them use it! It will save time, and for patients with inhibitors the factor they bring may be the only one that works for them!

18. Do not waste factor, it is $$$$; use the closest vial size to the recommended dose, and give the whole vial. Giving too much factor will ensure it lasts longer.

19. Ask the patient or parents questions, they will likely know more than you about their illness and can answer questions.

20. Factor needs to be given in its own line, it can not be given with anything else! Typically, you will use a butterfly needle to give it.
A 13 yo M presents with bleeding gums, he states that he has intermittent nose bleeds, and that he believes his family has an unknown bleeding disorder. Which of the following disorders is most likely?

**von Willebrand’s disease (vWD)** is the most common inherited bleeding disorder, with a prevalence of roughly 1/100. With vWD disease, platelets are normal in number, morphology, and other functions, but their adhering properties are diminished. Classic symptoms of vWF include easy bruising, gingival bleeding, epistaxis, hematuria, heavy menses, hematomas and hemarthroses. Laboratory values are not always useful for diagnosis, prothrombin time (PT), partial thromboplastin time (PTT), platelets and wWF levels can all be normal; bleeding time is occasionally prolonged. Treatment of bleeding in vWD includes desmopressin acetate (DDAVP), factor VIII, cryoprecipitate, antifibrinolytics, estrogens and topical agents such as fibrin glue or thrombin spray.

35 y/o M with hemophilia A presents with right knee pain. He says that he believes he is bleeding in his knee. He weighs 80 kg, and believes his factor level is <2%. How much factor must be used to reach 40% activity?

**1600U**

Remember: Factor VIII = Each U/kg = an Increase of Factor activity by **2%**
- 25 U/kg = 50% correction (for minor bleeds)
- 50 U/kg = 100% correction (for major bleeds)

GOAL 40% - 0% Activity Level = 40%
80 kg x 40% x 50U/kg = 80 x .4 x 50 = 1600

A 12 y/o M with hemophilia B presents with complaints of getting hit in the flank by a basketball just prior to arrival. He states that he has severe pain on that side. He doesn’t know his factor level. He weighs 35kg. How much factor should the patient receive?

**3500U**

Remember: Factor IX = Each U/kg = an Increase of Factor activity by **1%**
- 50 U/kg = 50% correction (for minor bleeds)
- 100 U/kg = 100% correction (for severe bleeds)

GOAL 100% - 0% Activity = 100% change
35kg x 100% x 100U/kg = 35 x 1 x 100 = 3500
A 22 y/o M with Hemophilia A presents with complaints of gum bleeding x 2 hours. He states that he was unable to give himself factor at home but is concerned. He states that he has a factor level of 5% and requests that you raise his level to 50%. He states that he weighs 75kg.

1688U

Remember: Factor VIII = Each U/kg = an Increase of Factor activity by 2%
  25 U/kg = 50% correction (for minor bleeds)
  50 U/kg = 100% correction (for major bleeds)

GOAL 50% - 5% Activity Level= 45%
75 kg x 45% x 50U/kg = 75 x .45 x 50 = 1688
BONUS: ACQUIRED HEMOPHILIA Information

These are patients with inhibitors to clotting factors making treatment more difficult.

Usually Caused by:
5. Malignancies (CLL, Adenocarcinomas)
6. Pregnancy or Postpartum State
7. Autoimmune Disorders (SLE, RA)
8. Idiopathic

If you are unsure if the patient has Factor Inhibitors, you can perform the Clotting Factor Mixing Test
Mix normal plasma with the patients serum
If the patient is Normal and does not have Acquired Hemophilia= aPTT will normalize
If the patient has Acquired Hemophilia or Factor Inhibitors = aPTT remains prolonged

Low titers of inhibitor: treat with Factor VIII concentrates

High titers of inhibitor: treat with factor VIIa concentrates or prothrombin complexes (PCCs)

TREATMENT FOR PATIENTS WITH INHIBITORS:

Recombinant Factor VIIa: “bypasses” the inhibitors.
Dose: 90-150 mcg/kg
Has a short half-life and may require multiple additional doses

Factor Eight Inhibitor Bypassing Agent: is a plasma-derived bypassing agent.
Dose: 50 U/kg
Caution: Must be given slowly, because rapid infusion can cause excessive clotting (e.g. stroke, limb ischemia, etc).
REFERENCES & READING:


