

The EM Educator Series

Mini-Case: Bleeding until proven otherwise...

Author: Alex Koyfman, MD (@EMHighAK) // Edited by: Brit Long, MD (@long_brit) and Manpreet Singh, MD (@MprizzleER)

A 12-year-old male is brought in by EMS after a fall from the monkey bars at school. He hit his head, and upon presentation, appears altered. He has a significant hematoma. His mother rushes into the room and tells you he has a history of hemophilia A. Your heart skips a beat with this new information.

Questions for Learners:

- 1) What are the different types of hemophilia?
- 2) How do these patients present, and what do you need to consider in evaluating/managing these patients?
- 3) ED work-up; don't wait to return!!
- 4) How do you provide factor replacement (bleeding site dependent)?
- 5) How can you use your cognitive approach to high-risk, low prevalence diseases (such as hemophilia) and apply these concepts and lessons learned elsewhere?

Suggested Resources:

- ✓ FOAMed:
 - <http://rebelem.com/hemophilia-whats-bloody-funny/>
 - <https://wikem.org/wiki/Hemophilia>
 - <https://emsimcases.com/tag/hemophilia/>
 - <http://pedemmorsels.com/hemophilia-ed/>
 - <https://www.orthobullets.com/pediatrics/4113/hemophilia>
- ✓ Articles:
 - <https://www.ncbi.nlm.nih.gov/pubmed/29277492>
 - <https://www.ncbi.nlm.nih.gov/pubmed/17976772>
- ✓ Podcasts:
 - <http://www.pemed.org/blog/2015/5/28/hemophillia>

Answers for Learners:

1) What are the different types of hemophilia?

“Hemophilia A (Classic Hemophilia)

- Deficiency in Factor VIII
- ≈80% of Hemophilia Cases (Factor VIII has 186,000 base pairs and therefore more prone to mutations than Factor IX)
- 1 in 5,000 male births

Hemophilia B (Christmas Disease)

- Deficiency in Factor IX
- ≈20% of Hemophilia Cases (Factor IX has 34,000 base pairs and therefore less prone to mutations than Factor VIII)
- 1 in 30,000 male births

Acquired Hemophilia

- These are patients with inhibitors to clotting factors making treatment more difficult.
- Causes:
 - Malignancies (CLL, Adenocarcinomas)
 - Pregnancy or Postpartum State
 - Autoimmune Disorders (SLE, RA)
 - Idiopathic
- Clotting Factor Mixing Test
 - Mix normal plasma with patients serum
 - Normal = aPTT will normalize
 - Inhibitor = aPTT remains prolonged
- Low titers of inhibitor (≤5 Bethesda Units) can still be treated with Factor VIII concentrates
- High titers of inhibitor (>5 Bethesda Units) may require treatment with factor VIIa concentrates or prothrombin complexes (PCCs)” via R.E.B.E.L. EM

2) How do these patients present, and what do you need to consider in evaluating/managing these patients?

“The clinical manifestations of hemophilia A and B are practically indistinguishable and characterized primarily by easy bruising and recurrent bleeding into joints and deep muscles. Hemarthrosis is the most common complication of hemophilia with elbows, knees, and ankles most often affected. An important point to emphasize is patients may develop pain in the affected joint before any clinical signs can be detected, so it is important that even vague joint symptoms be taken seriously.

Intracranial hemorrhage (ICH) is the most common noninfectious cause of death in hemophiliacs. A history of trauma may be part of the presentation but severe hemophiliacs are at high risk for atraumatic ICH. Any patient with known hemophilia presenting with a history of or suspected head trauma or signs/symptoms suspicious for ICH should be assumed to be bleeding and managed aggressively with consideration of emergent factor replacement and possible admission for observation and neuro checks.” via R.E.B.E.L. EM

3) ED work-up; don't wait to return!!

Work-Up

- Coags
 - Only helpful in making the diagnosis; once established unlikely to yield new information
 - PT - normal
 - PTT - abnormal (unless mild hemophilia)
 - PTT s/p factor - should correct to normal
- Factor VIII assay

Consider

- Head CT
 - If headache, altered mental status, significant blunt head injury
 - MRI is preferred to CT if available (better visualization of posterior fossa)
- CT A/P
 - Back, thigh, groin, or abdominal pain
- [LP](#)
 - Replete factor before attempting

Evaluation

- Pain in soft tissue is bleeding until proven otherwise
- Paresthesias in legs - consider retroperitoneal bleed
- Flexion contracture at the hip- iliopsoas bleed
- Easy bruising or bleeding out of proportion to the history of trauma
- Recurrent bleeding into joints and muscles
 - Arthrocentesis not indicated
- Prolonged PTT; normal PT

Factor VIII assay

Consider before treatment (for heme to follow)

- Normal: 50-150%
- Mild: >5% (usually an insult causes bleeding)
- Moderate: 1-5% (usually an insult causes bleeding)
- Severe: <1% (spontaneous bleeding not uncommon, multiple bleeding episodes/month)

4) How do you provide factor replacement (bleeding site dependent)?

Factor Replacement

- Major bleeding (GI, CNS, large muscle, trauma) requires factor replacement level 80-100%
- Moderate bleeding (soft tissue, small muscle, joint) requires 30-50%
- Diagnosis unknown
 - Give FFP (contains VIII and IX)
 - Each bag raises factor levels by 3-5%
- Severe mechanism of injury (head, neck trauma), even without evidence of bleeding
 - Hemophilia is factor deficiency, not platelet deficiency/malfunction so initial hemostasis may be achieved but clot stabilization will not persist
 - Delayed bleeding is a serious risk, so factor replacement must occur immediately

5) How can you use your cognitive approach to high-risk, low prevalence diseases (such as hemophilia) and apply these concepts and lessons learned elsewhere?

Have a high sensitivity for these conditions by having a high index of suspicion based on the clinical picture. Prove to yourself why they do not have that condition....deduction!