



**Rap #27, Heme/ Onc**

**Dangers of Infections in Splenectomy Patients (EMDocs, Authored by Dilani Weerasuriya, MD) - Review by Steve Palm, MD**

What infections are splenectomy patients most at risk for?

Encapsulated organisms	Streptococcus pneumoniae <sup>1</sup> <ul style="list-style-type: none"><li>- The most common cause of infection and death in asplenic patients<sup>3</sup></li></ul> Neisseria meningitides <sup>2</sup> Haemophilus influenza <sup>2</sup>
Gram negative rods	Capnocytophaga canimorsus <sup>1</sup> <ul style="list-style-type: none"><li>- Especially if scratched or bitten by a dog</li></ul>
Parasitic infections	Malaria Babesiosis <sup>2</sup>

- The highest risk of infection is 90 days after splenectomy, but it remains significantly higher for the first 2 years following surgery.
- Streptococcus pneumoniae is still the most common cause of sepsis and death in these patients.
- Early broad spectrum antibiotics are important!
  - Mortality rates in post-splenectomy infection range from 50% to 70%.
  - Do not delay antibiotics to perform diagnostic testing. Asplenic patients can decompensate quickly, therefore any possible infection should be treated with antibiotics rapidly. Their mortality rate may be decreased from 70% to 10% with effective and early management.

Tier 1: BEM Rater Scale	Score-choose only 1	Tier 2: Content accuracy	Score-choose only 1	Tier 3: Educational Utility	Score-choose only 1	Tier 4: EBM	Score-choose only 1	Tier 5: Referenced	Score-choose only 1
Assuming that the results of this article are valid, how much does this article impact on EM clinical practice?		Do you have any concerns about the accuracy of the data presented or conclusions of this article?		Are there useful educational pearls in this article for residents?		Is this article reflect evidence based medicine (EBM) and thus lack bias?		Are the authors and literature clearly cited?	
Useless information	<input type="radio"/>	Yes, many concerns from many inaccuracies	<input type="radio"/>	Low value: No valuable pearls	<input type="radio"/>	Not EBM based, only expert opinion (and thus more biased)	<input type="radio"/>	No	<input type="radio"/>
Not really interesting, not really new, changes nothing	<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>
Interesting and new, but doesn't change practice	<input checked="" type="radio"/>	Yes, a major concern about few inaccuracies	<input type="radio"/>	Yes, but there are only a few (1-2) valuable or multiple (>=3) less-valuable educational pearls	<input checked="" type="radio"/>	Minimally EBM based	<input type="radio"/>		<input type="radio"/>
Interesting and new, has the potential to change practice	<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>
New and important: this would probably change practice for some EPs	<input type="radio"/>	Minimal concerns over minor inaccuracies	<input type="radio"/>	Yes, there are several (>=3) valuable educational pearls, or a few (1-2) KEY educational pearls that every resident should know before graduating	<input type="radio"/>	Mostly EBM based	<input checked="" type="radio"/>		<input type="radio"/>
New and Important: this would change practice for most EPs	<input type="radio"/>		<input checked="" type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input checked="" type="radio"/>
This is a "must know" for EPs	<input type="radio"/>	No concerns over inaccuracies	<input type="radio"/>	Yes, there are multiple KEY educational pearls that residents should know before graduating	<input type="radio"/>	Yes exclusively EBM based (unbiased)	<input type="radio"/>	Yes	<input type="radio"/>
Your Score	3		6		3		5		6

**Rapid Review: Acute Intermittent Porphyria (First10EM, Authored by Justin Morgenstern, MD)- Reviewed by Hilary Royston, MD**

The First10EM blogpost was an overview of the article: *Bissell DM, Anderson KE, Bonkovsky HL. Porphyria. The New England journal of medicine. 2017; 377(9):862-872*

**Porphyria Overview:**

- Group of disorders involving enzymatic defects in heme biosynthetic pathway.
- 8 forms: usually separated into acute vs cutaneous.
- Manifests as neurologic and/or cutaneous symptoms.

**Acute Intermittent Porphyria:**

- Most common type encountered in ED
- Presentation:
  - Most commonly young healthy females
  - Nonspecific symptoms: abdominal pain, nausea, vomiting, extremity pain, muscle weakness, AMS, seizures (in up to 20%)
    - Triad: seizures, hyponatremia, non-specific abdominal pain in young female
  - Usually previous ED visits with non-diagnostic evaluation
  - Labs: possible minimal elevation LFT, hyponatremia
- Triggers:
  - Progesterone surge, caloric deprivation, OCPs
- Diagnosis:
  - **Elevated random urine or plasma porphobilinogen level**
- Treatment:
  - Benzos for seizures (antiepileptics can make things worse)
  - Supportive care, symptom control
  - IV heme (panhematin)
- Prognosis:
  - Previous mortality rate 25%, now better with improved diagnosis
  - If motor neuron symptoms are present → recovery can be up to 1 year

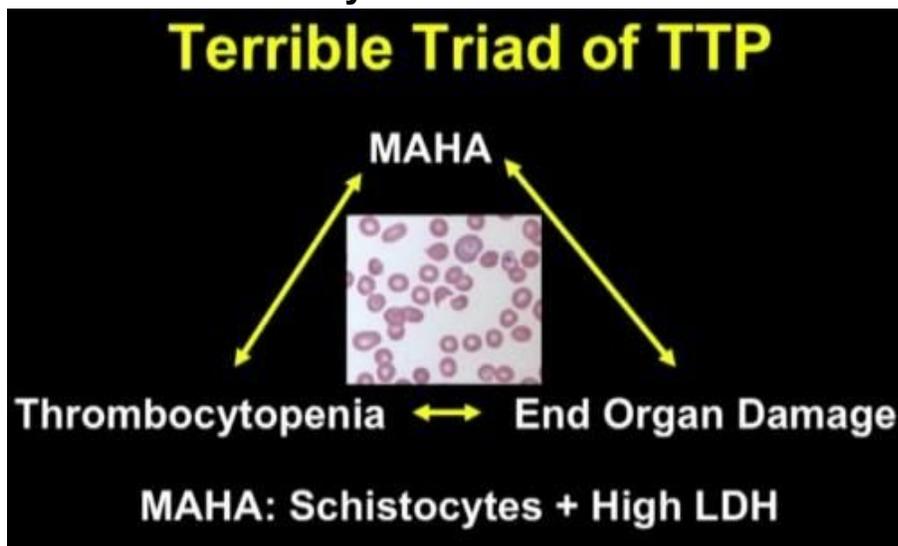
**FUN FACT: VAMPIRES:**

- Cutaneous porphyria ? origin vampire legend: skin changes following sun exposure, red/brown urine ? ingestion human blood

Tier 1: BEEM Rater Scale	Score-choose only 1	Tier 2: Content accuracy	Score-choose only 1	Tier 3: Educational Utility	Score-choose only 1	Tier 4: EBM	Score-choose only 1	Tier 5: Referenced	Score-choose only 1
Assuming that the results of this article are valid, how much does this article impact on EM clinical practice?		Do you have any concerns about the accuracy of the data presented or conclusions of this article?		Are there useful educational pearls in this article for residents?		Is this article reflect evidence based medicine (EBM) and thus lack bias?		Are the authors and literature clearly cited?	
Useless information	<input type="radio"/>	Yes, many concerns from many inaccuracies	<input type="radio"/>	Low value: No valuable pearls	<input type="radio"/>	Not EBM based, only expert opinion (and thus more biased)	<input type="radio"/>	No	<input type="radio"/>
Not really interesting, not really new, changes nothing	<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>
Interesting and new, but doesn't change practice	<input type="radio"/>	Yes, a major concern about few inaccuracies	<input type="radio"/>	Yes, but there are only a few (1-2) valuable or multiple (>=3) less-valuable educational pearls	<input type="radio"/>	Minimally EBM based	<input type="radio"/>		<input type="radio"/>
Interesting and new, has the potential to change practice	<input checked="" type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input checked="" type="radio"/>		<input type="radio"/>
New and important: this would probably change practice for some EPs	<input type="radio"/>	Minimal concerns over minor inaccuracies	<input type="radio"/>	Yes, there are several (>=3) valuable educational pearls, or a few (1-2) KEY educational pearls that every resident should know before graduating	<input checked="" type="radio"/>	Mostly EBM based	<input type="radio"/>		<input type="radio"/>
New and Important: this would change practice for most EPs	<input type="radio"/>		<input checked="" type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input checked="" type="radio"/>
This is a "must know" for EPs	<input type="radio"/>	No concerns over inaccuracies	<input type="radio"/>	Yes, there are multiple KEY educational pearls that residents should know before graduating	<input type="radio"/>	Yes exclusively EBM based (unbiased)	<input type="radio"/>	Yes	<input type="radio"/>
Your Score	4		6		5		4		6

**TTP/ DIC Part I: Diagnosis** (EMCrit, Authored by Tom DeLoughery, MD) - Review by Enyioma Okechukwu, MD

- **Thrombotic Thrombocytopenic Purpura (TTP):** A primary disease of platelets
  - **Pathophysiology:**
    - Platelets begin to aggregate in vessels within organs, which explain clinical presentation that correspond to which organ it is deposited in. Most common cause of death in TTP is cardiac. You can also see renal sx, neuro sx, ARDS, pancreatitis, etc. Because it can have a widespread range of presentations, must always keep on the differential.
    - This is caused by a deficiency of ADAM-TS-13, which is a proteinase that cleaves vonWillibrand Factor (vWF). Without vWF the result is large molecules that lead to them clumping together. Can be genetic or acquired.
      - This lab level takes too long to come back so TTP is a clinical diagnosis
  - **Diagnosis:** LDH is extremely high up to 4-5x normal; **No schistocytes = Not TTP**



- **Disseminated Intravascular Coagulation (DIC):** disease of thrombin generation
  - **Pathophysiology:**

- Increased thrombin generation, which normally has roles in thrombosis (leads to fibrin to bind together) as well as fibrinolysis (from feedback). So overproduction results in powerful stimulus for coagulation, leading to factor destruction and this causes patients clot and bleed.
- Secondary to some other disease process. Anything that generates too much thrombin can result in DIC.
  - Examples include sepsis, OB disasters, trauma, snake bites, etc.
  - Anything that disrupts endothelium and triggers coagulation cascade
- **Diagnosis:**
  - Increase in all of your coagulation tests INR, aPTT, PTT, and very high D-dimer; meanwhile you have dropping platelets and fibrinogen
  - D-dimer is extremely elevated (often > 15-20k)

IG: @hansonzanatomy

# THROMBOCYTOPENIA

## TTP

THROMBOTIC THROMBOCYTOPENIA PURPURA

CONGENITAL OR ACQUIRED  
**ADAMTS13**  
DEFICIENCY = GIANT vWF  
CAUSING PLATELET AGGREGATION  
& FORMATION OF **HYALINE THROMBI**  
IN CAPILLARIES & ARTERIOLES

## DIC

DISSEMINATED INTRAVASCULAR COAGULATION

ACQUIRED  
COAGULATION DISORDER  
CAUSING **FIBRIN THROMBI**  
TO DEPOSIT IN SMALL  
VESSELS, WHILE  
PLATELETS & CLOTTING  
FACTORS ARE RAPIDLY  
CONSUMED

### 1

1<sup>o</sup> DUE TO PRIMARY ENZYME DEFICIENCY

PETECCHIAE (COALESCE TO FORM PURPURA)

HEMOLYTIC ANEMIA

THROMBOCYTOPENIA

NEURO SXS

RENAL DYSFXN

END ORGAN DAMAGE

BOTH PATIENTS CAN LOOK VERY, VERY, VERY SICK

### 2

2<sup>o</sup> DUE TO OTHER DISEASE PROCESS

PURPURA

SCHISTOCYTES

## Tx: PLASMA EXCHANGE

EXCHANGE BLOOD VOLUME x 1.5 FOR ABOUT 5 DAYS

- FRESH FROZEN PLASMA → 2 UNIT BOLUS + 1 UNIT q6hr
- STEROIDS (60mg PREDNISONE OR 125mg SOLUMEDROL)

NEVER GIVE PLATELETS!!!

## TREAT PRIMARY CAUSE: Tx

& CORRECT COAGULATION ABNORMALITIES

- PLATELETS > 50,000
- FIBRINOGEN > 150 (>200 IF OB)
- HCT > 21%
- INR < 2-3
- aPTT < 1.5x NML

GIVE CRYO (100 UNITS)

Tier 1: BEEM Rater Scale	Score-choose only 1	Tier 2: Content accuracy	Score-choose only 1	Tier 3: Educational Utility	Score-choose only 1	Tier 4: EBM	Score-choose only 1	Tier 5: Referenced	Score-choose only 1
Assuming that the results of this article are valid, how much does this article impact on EM clinical practice?		Do you have any concerns about the accuracy of the data presented or conclusions of this article?		Are there useful educational pearls in this article for residents?		Is this article reflect evidence based medicine (EBM) and thus lack bias?		Are the authors and literature clearly cited?	
Useless information	<input type="radio"/>	Yes, many concerns from many inaccuracies	<input type="radio"/>	Low value: No valuable pearls	<input type="radio"/>	Not EBM based, only expert opinion (and thus more biased)	<input type="radio"/>	No	<input checked="" type="radio"/>
Not really interesting, not really new, changes nothing	<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>
Interesting and new, but doesn't change practice	<input type="radio"/>	Yes, a major concern about few inaccuracies	<input type="radio"/>	Yes, but there are only a few (1-2) valuable or multiple (>=3) less-valuable educational pearls	<input type="radio"/>	Minimally EBM based	<input type="radio"/>		<input type="radio"/>
Interesting and new, has the potential to change practice	<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>
New and important: this would probably change practice for some EPs	<input checked="" type="radio"/>	Minimal concerns over minor inaccuracies	<input type="radio"/>	Yes, there are several (>=3) valuable educational pearls, or a few (1-2) KEY educational pearls that every resident should know before graduating	<input type="radio"/>	Mostly EBM based	<input checked="" type="radio"/>		<input type="radio"/>
New and Important: this would change practice for most EPs	<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>
This is a "must know" for EPs	<input type="radio"/>	No concerns over inaccuracies	<input checked="" type="radio"/>	Yes, there are multiple KEY educational pearls that residents should know before graduating	<input checked="" type="radio"/>	Yes exclusively EBM based (unbiased)	<input type="radio"/>	Yes	<input type="radio"/>
Your Score	5		7		7		5		1

**TTP/ DIC Part II: Treatment** (EMCrit, Authored by Tom DeLoughery, MD) - Review by Andrea Weiers, MD

**TTP:**



- Don't feed the fire
- Disease of overactive platelets → spontaneously aggregate
  - **Never give platelets**, makes things worse due to this spontaneous aggregation.
  - **This is feeding the fire.** Patients are usually hypercoagulable even with low platelet count.
- Plasma exchange
  - Multiple advantages
    - Remove inhibitors of ADAMS-TS13
    - Replace with plasma containing ADAMS-TS13
    - Effective way of giving fluids, get 13-14 units of platelets
  - May need multiple treatments
  - Requires a dialysis catheter
    - Don't shy away because platelets are low. Risk of bleed is very low.
    - Consider placing femorally for ability to apply pressure.
  - While waiting (for line, techs, transfer):
    - FFP: has ADAMS-TS13, also slows spontaneous platelet aggregation
      - Infuse 1-2 off the bat, then 1U q6H
    - Steroids: TTP usually autoimmune
      - Dose is arbitrary, he uses prednisone 60 mg, others use methylpred 125 mg
- Untreated mortality 90-100% → 20% with plasma exchange

**DIC:**

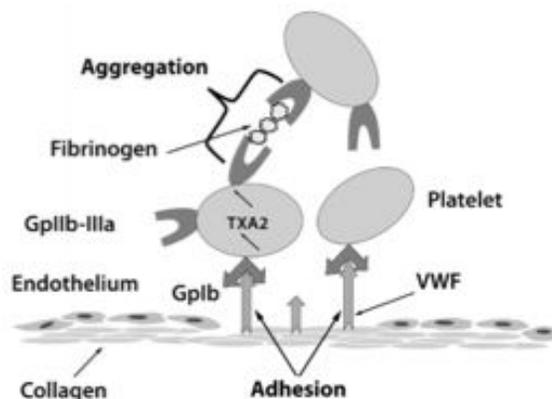
- Don't worry about "feeding the fire"
- **Treat underlying cause** e.g. sepsis
- Support coagulation (patients are bleeding), especially if needing a procedure
  - Magic 5 ("trauma coag panel"), based on 5 tests of bleeding/coagulation stability
    - Fibrinogen >150 mg/dL (>200 mg/dL if OB disaster)

- Key for clot strength
- Use cryoprecipitate, usually 10U
- Platelets >50,000
- Hct >21% (don't go too crazy)
- aPTT <1.5x normal
  - Use plasma – replaces coagulation factors, inhibitors of fibrinogen, and inhibitors of coagulation
- INR <2-3
  - He doesn't recommend treatment until this is 2+ because many ill people will have INR up to 1.5
- As this is a disease of too much thrombin... Why don't we inhibit thrombin with heparin??
  - Studies show terrible outcome
  - Doesn't address thrombocytopenia, other coag abnormalities
  - Only uses if macrothrombosis e.g. massive PE in cancer patient
- What about TXA? Nope.
  - DIC = excess thrombin generation and excess thrombinolysis
  - If you blocked the thrombinolysis with TXA → even more prothrombotic state

Tier 1: BEEM Rater Scale	Score-choose only 1	Tier 2: Content accuracy	Score-choose only 1	Tier 3: Educational Utility	Score-choose only 1	Tier 4: EBM	Score-choose only 1	Tier 5: Referenced	Score-choose only 1
Assuming that the results of this article are valid, how much does this article impact on EM clinical practice?		Do you have any concerns about the accuracy of the data presented or conclusions of this article?		Are there useful educational pearls in this article for residents?		Is this article reflect evidence based medicine (EBM) and thus lack bias?		Are the authors and literature clearly cited?	
Useless information	<input type="radio"/>	Yes, many concerns from many inaccuracies	<input type="radio"/>	Low value: No valuable pearls	<input type="radio"/>	Not EBM based, only expert opinion (and thus more biased)	<input type="radio"/>	No	<input type="radio"/>
Not really interesting, not really new, changes nothing	<input checked="" type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input checked="" type="radio"/>
Interesting and new, but doesn't change practice	<input type="radio"/>	Yes, a major concern about few inaccuracies	<input type="radio"/>	Yes, but there are only a few (1-2) valuable or multiple (>=3) less-valuable educational pearls	<input checked="" type="radio"/>	Minimally EBM based	<input checked="" type="radio"/>		<input type="radio"/>
Interesting and new, has the potential to change practice	<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>
New and important: this would probably change practice for some EPs	<input type="radio"/>	Minimal concerns over minor inaccuracies	<input type="radio"/>	Yes, there are several (>=3) valuable educational pearls, or a few (1-2) KEY educational pearls that every resident should know before graduating	<input type="radio"/>	Mostly EBM based	<input type="radio"/>		<input type="radio"/>
New and Important: this would change practice for most EPs	<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>
This is a "must know" for EPs	<input type="radio"/>	No concerns over inaccuracies	<input checked="" type="radio"/>	Yes, there are multiple KEY educational pearls that residents should know before graduating	<input type="radio"/>	Yes exclusively EBM based (unbiased)	<input type="radio"/>	Yes	<input type="radio"/>
Your Score	2		7		3		3		2

## Platelets and ddAVP in the Management of Intracranial Hemorrhage (EMDocs, Authored by Jeremy Kim, MD - Review by Mary Swiggum, MD)

- Are patients taking anti-platelet agents at higher risk of developing ICH?
  - Short answer is yes; even patients taking aspirin (particularly moderate to high doses) are at increased risk as well.
- Biochem review
  - Platelets go to sites of endothelial damage and bind by vWf and Gp1b
  - COX-1, TXA2, ADP, GPIIb-IIIa further recruit plts/help aggregation
  - Aspirin = irreversible COX-1 inhibitor, rapidly metabolized (short duration of effect)
  - Clopidogrel = ADP receptor inhibitors. Active metabolites, lasts 5-7 days
  - **ddAVP: promotes vWF production**



- Concern is that patients on anti-platelet medications will have worsening bleeds and clinical outcomes. Literature does not consistently show this.
- Current literature not conclusive on management
- Article suggests consider giving platelets and ddAVP (0.3 mcg/kg)

Tier 1: BEEM Rater Scale	Score-choose only 1	Tier 2: Content accuracy	Score-choose only 1	Tier 3: Educational Utility	Score-choose only 1	Tier 4: EBM	Score-choose only 1	Tier 5: Referenced	Score-choose only 1
Assuming that the results of this article are valid, how much does this article impact on EM clinical practice?		Do you have any concerns about the accuracy of the data presented or conclusions of this article?		Are there useful educational pearls in this article for residents?		Is this article reflect evidence based medicine (EBM) and thus lack bias?		Are the authors and literature clearly cited?	
Useless information	<input type="radio"/>	Yes, many concerns from many inaccuracies	<input type="radio"/>	Low value: No valuable pearls	<input type="radio"/>	Not EBM based, only expert opinion (and thus more biased)	<input type="radio"/>	No	<input type="radio"/>
Not really interesting, not really new, changes nothing	<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>
Interesting and new, but doesn't change practice	<input checked="" type="radio"/>	Yes, a major concern about few inaccuracies	<input type="radio"/>	Yes, but there are only a few (1-2) valuable or multiple (>=3) less-valuable educational pearls	<input checked="" type="radio"/>	Minimally EBM based	<input type="radio"/>		<input type="radio"/>
Interesting and new, has the potential to change practice	<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>
New and Important: this would probably change practice for some EPs	<input type="radio"/>	Minimal concerns over minor inaccuracies	<input checked="" type="radio"/>	Yes, there are several (>=3) valuable educational pearls, or a few (1-2) KEY educational pearls that every resident should know before graduating	<input type="radio"/>	Mostly EBM based	<input checked="" type="radio"/>		<input type="radio"/>
New and Important: this would change practice for most EPs	<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>		<input type="radio"/>
This is a "must know" for EPs	<input type="radio"/>	No concerns over inaccuracies	<input type="radio"/>	Yes, there are multiple KEY educational pearls that residents should know before graduating	<input type="radio"/>	Yes exclusively EBM based (unbiased)	<input type="radio"/>	Yes	<input checked="" type="radio"/>
<b>Your Score</b>	<b>3</b>		<b>5</b>		<b>3</b>		<b>5</b>		<b>7</b>

**Summary edited by Joe Walter, Zach Finn, Sara Schroeder, Ollie Garrison and Dilian Stoimenova**