Go With the Flow

Hematologic Emergencies

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Blood Circulating Tissue
7% of body weight
Adult volume = 5 liters
Surface area of erythrocytes 2000 times the body's exterior surface

Components of Blood
- Plasma
- Cells
  - Red blood cells (96% of formed elements)
  - White blood cells (3%)
  - Platelets (1%)
- Trace elements
  - Albumin, clotting factors, immunoglobulins
  - Hormones, electrolytes

Functions of Blood
- Delivery of nutrients (O2, glucose)
- Removal of waste products (CO2, lactic acid)
- Transport of cells between tissues and organs
- Mobile immune system
- Delivers coagulation materials
- Transfers heat to skin
- Buffers pH of body
Structure of RBC’s

- Anucleate when mature,
- Shape = biconcave disc
- Human RBC = 6-8 micrometers
- Total RBC surface area = 3000 sq. miles
- 2 million produced every second

Red Blood Cell Issues

- Too few
- Too many
- Wrong shape
- Impaired Function
Anemia

- Laboratory Assessment
  - Hematocrit (packed cell volume)
  - Reticulocyte count
  - Red cell indices
    - Mean cell volume (MCV)
    - Mean corpuscular hemoglobin (MCH)
    - Mean cell hemoglobin concentration (MCHC)

Peripheral Smear

- Critical to making the diagnosis
- You have to request it
- You can find
  - Inclusions, stippling, spherocytes, sickling, rouleaux formations, blister cells, hypochromia, hyperchromia, fragmentation, infections, WBC abnormalities, platelet abnormalities

Mnemonic

<table>
<thead>
<tr>
<th>H</th>
<th>History</th>
</tr>
</thead>
<tbody>
<tr>
<td>E</td>
<td>Etiology--congenital vs. acquired</td>
</tr>
<tr>
<td>M</td>
<td>Medications</td>
</tr>
<tr>
<td>E</td>
<td>Exam</td>
</tr>
<tr>
<td>P</td>
<td>Peripheral Smear</td>
</tr>
<tr>
<td>A</td>
<td>Additional diagnostics</td>
</tr>
<tr>
<td>T</td>
<td>Transfusion / treatment</td>
</tr>
<tr>
<td>H</td>
<td>Help</td>
</tr>
</tbody>
</table>
Threshold for intervention

<table>
<thead>
<tr>
<th>Element</th>
<th>Less than</th>
<th>More than</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hgb</td>
<td>7.5 g/dl</td>
<td>20 g/dl</td>
</tr>
<tr>
<td>ANC</td>
<td>500 / µl</td>
<td>blasts 200K</td>
</tr>
<tr>
<td>Platelets</td>
<td>10,000/µl</td>
<td>1,000,000/µl</td>
</tr>
</tbody>
</table>

Factor deficiency and platelet dysfunction is treated if clinically significant regardless of numerical level.

Case 1

- H: 35 BM, fever, cough, CP x 2 days
- E: h/o Sickle Cell
- M: None
- T: T 101.1, HR 132, RLL ↓ BS
- P: WBC 13.8, Hct 24.1, Plt 288, smear
- A: Additional Studies?
- T: Treatment?
- H: ID, Ortho, Urology?, Heme

Case 1 Smear
Sickle Cell Acute Chest Syndrome

1. Manage vasoocclusion
   - Hydration and oxygen

2. Manage pain
   - Morphine and analogs
   - Avoid meperidine (demerol)

3. Manage hemolytic anemia
   - Folate depleted
   - Check for iron deficiency
   - Ensure brisk reticulocyte response

4. Treat infections

5. Manage end-organ damage
   - Transfusion to ↓ HbS to 30%

6. Prevent stroke

7. Treat pulmonary hypertension
Case 2

**History**
- 42 BM, s/p meth, MRSA, fatigue, SOB

**Exam**
- Acute

**Medications**
- HCTZ, prozac, prilosec, TMP, hibiclens

**PF**
- AF, P = 132, BP 105/75, exam nonfocal

**Lab**
- Hct 17, RBC indices nl, plt 175, dif nl, smear

**Additional studies?**

**Treatment?**

**Help?**

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**Hemolytic Anemia**

- Over 200 different causes
- This case is G-6-PD (glucose-6-phosphatase dehydrogenase) deficiency exacerbated by TMP
- Diagnosis made by smear--Heinz bodies
- D/C the TMP and avoid oxidative medications
- Transfuse
**Case 3**

<table>
<thead>
<tr>
<th>H</th>
<th>42 y/o WF, HA, confusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>E</td>
<td>Acute</td>
</tr>
<tr>
<td>M</td>
<td>None</td>
</tr>
<tr>
<td>E</td>
<td>T = 101.1, P = 115, pallor, petechiae</td>
</tr>
<tr>
<td>P</td>
<td>Hgb 7.2, Platelets 45K, BUN 30, Cr. 2.4</td>
</tr>
<tr>
<td>A</td>
<td>Additional studies?</td>
</tr>
<tr>
<td>T</td>
<td>Treatment options?</td>
</tr>
<tr>
<td>H</td>
<td>Help?</td>
</tr>
</tbody>
</table>

**Peripheral Smear Case 3**

**Thrombotic Thrombocytopenic Purpura**
- Pentad of microangiopathic hemolytic anemia, thrombocytopenia, neurological symptoms, fever, renal dysfunction (40%)
- Viral prodrome
- Associated with pregnancy, HIV, and cancers
- True medical emergency
- Patients treated with plasma exchange / plasmapheresis
White Blood Cells

- Produced in bone marrow
- Integral part of immune system
- Different types = different functions
  - Neutrophils—phagocytosis of bacteria
  - Eosinophils—viral and parasites
  - Basophils—histamine, inflammation
  - Macrophages—phagocytes

Phagocytosis

WBC problems

- Too many
- Too few
- Not enough diversity
- Impaired function
Case 4

H 59 y/o male, fever, sore throat
E Acute
M Lisinopril, HCTZ, Potassium
E T 102, P 115, R 15, BP 135/87, SaO2 95% RA
P WBC 200K / µl, Hgb 9.2, platelets 35K
A additional studies?
T Treatment?
H Help?

Smear Case 4

Strep pharyngitis
- Underlying diagnosis = Acute Mylogenous Leukemia
- Often presents as an infection
- Must r/o pneumonias
Case 5

H 29 year old, fever, fatigue, 14 d s/p chemo
E Acute
M Reglan, ensure
E T 102.5, P 100, R 14, BP 115/75, SaO2 99% RA
P See next page
A additional studies?
T Treatment?
H Help?

CBC

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Value</th>
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<tbody>
<tr>
<td>WBC</td>
<td>2.2 x 1000/µl</td>
</tr>
<tr>
<td>Hct</td>
<td>38 G/dl</td>
</tr>
<tr>
<td>Lymph</td>
<td>0.54</td>
</tr>
<tr>
<td>Mid</td>
<td>0.31</td>
</tr>
<tr>
<td>Neutrophil</td>
<td>0.15</td>
</tr>
<tr>
<td>Platelets</td>
<td>178K</td>
</tr>
</tbody>
</table>

Febrile Neutropenia

- ANC < 500/µl
- Medical emergency
- Progresses rapidly
- Transfer to ED!!!
Intermission

Glue
- Clotting mechanism
- Platelets
- Clotting cascade

Platelets
Case 6

<table>
<thead>
<tr>
<th>H</th>
<th>26 y/o male with tongue tenderness, flu 2 weeks</th>
</tr>
</thead>
<tbody>
<tr>
<td>E</td>
<td>Acute</td>
</tr>
<tr>
<td>M</td>
<td>OTC’s</td>
</tr>
<tr>
<td>E</td>
<td>VS normal, see next page</td>
</tr>
<tr>
<td>P</td>
<td>results?</td>
</tr>
<tr>
<td>A</td>
<td>additional studies?</td>
</tr>
<tr>
<td>T</td>
<td>Treatment?</td>
</tr>
<tr>
<td>H</td>
<td>Help?</td>
</tr>
</tbody>
</table>
Tongue

Pharynx

Thrombocytopenia

- Decreased number vs. defective platelets
- Thrombocytopenia
  - increased destruction
  - decreased production
- DDX = von Willebrand, drugs, uremia, bone marrow failure, infectious, malignancy, DIC, TB
- This case = HIV seroconversion

This case = HIV seroconversion
**Idiopathic Thrombocytopenic Purpura**
- adults = chronic condition
- Look for s/s of bleeding--purpura, petechiae, gingival bleeding, CNS
- Get the smear!
- Platelet transfusion for severe bleeding
- Rule change: adults with plt < 50,000 and bleeding need treatment
- Treatment = steroids and IV Ig

**Heparin-Induced Thrombocytopenia**
- Antibody-mediated reaction
- Onset may be delayed up to 3 weeks
- Rapid at re-exposure
- Mortality = 20%
- Limb amputations = 10%

**Formation of a Hemostatic Plug**
- Primary Hemostasis
  - Platelet adhesion
  - Platelet aggregation
  - Release of adenosine diphosphate
  - Activation of the coagulation cascade
- Secondary Hemostasis
vonWillebrand’s Disease
- vonWillebrand’s factor forms the bridge between vessel wall and platelet
- autosomal dominant
- 3 levels
  - 1 = 70% of population, mild disease
  - 2 = dysfunctional adherence, moderate bleeding
  - 3 = severe bleeding, like hemophilia
- Treatment with DDAVP, Factor VIII, cryoprecipitate

Clotting Cascade
A good model should also be complicated enough to reflect the realities of the biological system.
Hemophilia

Prevalance 1:5000 male births
Factor VIII—most common—Hemophilia A
Factor IX (Christmas Disease or Hemophilia B)
Graded based on amount of factor present
Most common cause of death of a hemophiliac is AIDS
Acquired hemophilia—no family history

Hemophilia Emergencies
Head injuries—treated as CNS bleeds
Throat/neck—any swelling in throat and neck must be evaluated as a bleed
Intramuscular—compartment syndrome or extensive bleed
Iliopsoas—bleed can mimic hip fracture and acute abdomen
### Case 7

<table>
<thead>
<tr>
<th><strong>H</strong></th>
<th>45 y/o male with sudden mental status changes</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>E</strong></td>
<td>Acute</td>
</tr>
<tr>
<td><strong>M</strong></td>
<td>None</td>
</tr>
<tr>
<td><strong>T</strong></td>
<td>T 101.9, HR 150, ronchi, petechiae, bleeding from needle sticks</td>
</tr>
<tr>
<td><strong>P</strong></td>
<td>WBC 30K, Hgb 9, platelets 20K</td>
</tr>
<tr>
<td><strong>A</strong></td>
<td>additional studies?</td>
</tr>
<tr>
<td><strong>T</strong></td>
<td>Treatment?</td>
</tr>
<tr>
<td><strong>H</strong></td>
<td>Help?</td>
</tr>
</tbody>
</table>

### Smear Case 7

![Blood smear with arrows indicating abnormal cells](image)

### Disseminated Intravascular Coagulation

- Always an underlying cause
- Systemic activation of coagulation system
- Coagulation proteins and platelets are consumed and exhausted
- Final common pathway for many acute conditions
Causes of DIC

<table>
<thead>
<tr>
<th>Causes</th>
<th>DIC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sepsis</td>
<td>OB</td>
</tr>
<tr>
<td>Trauma</td>
<td>OB</td>
</tr>
<tr>
<td>Pancreatitis</td>
<td>OB</td>
</tr>
<tr>
<td>Malignancy</td>
<td>OB</td>
</tr>
<tr>
<td>OB</td>
<td>OB</td>
</tr>
<tr>
<td>Vascular abnormalities</td>
<td>OB</td>
</tr>
<tr>
<td>Liver failure</td>
<td>OB</td>
</tr>
<tr>
<td>Recreational drugs</td>
<td>OB</td>
</tr>
<tr>
<td>Vector injuries</td>
<td>OB</td>
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<tr>
<td>DKA</td>
<td>OB</td>
</tr>
<tr>
<td>Fat embolism</td>
<td>OB</td>
</tr>
<tr>
<td>Transfusion</td>
<td>OB</td>
</tr>
</tbody>
</table>

Warfarin

- Impairs production of VII, IX, X, II, and Protein C and S which make up the intrinsic coagulation pathway
- Creation of these factors is controlled by Vitamin K
- Warfarin inhibits absorption of vitamin K therefore the production of factors

Warfarin “Overdose”

- PT / INR values in perspective
- Lab logistical issues
- Reversing anticoagulant effect
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