

EM Basic- Heme Onc Part 2- Hematology Emergencies

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Anemia in the ED

- Decreased volume of red blood cells
- Generally defined as hemoglobin <14 for males and <12 for females
 - Normal values are highly dependent on lab equipment
- Usual presentation (without obvious bleeding source)- sent in from outpatient office or symptoms of anemia- or found incidentally during ED workup

History

- Ask about symptoms of anemia- fatigue, weakness, dyspnea on exertion, chest pain
- Ask about blood in the stool or urine, ask menstruating females about a history of heavy or prolonged periods, ask about recent surgeries or GI procedures
- Past medical history? Renal disease? History of anemia or transfusions?
- Get rest of medical history- PMH, meds, allergies, medications, allergies, PSH

PEARL: Young patients may tolerate anemia down to a hemoglobin of 5 or 6, most have at least some symptoms at a hemoglobin of 7

Exam

- Skin, mucosal, or conjunctival pallor
- Jaundice (if having hemolytic anemia)
- Flow murmur on cardiac exam with severe anemia

Lab studies

- CBC- look for severity of anemia
- Mean Corpuscular Volume (MCV)
- General categories (not an exhaustive list but good enough for the ED workup)
 - Microcytic**- low MCV- usually iron deficiency anemia or anemia of chronic disease
 - Normocytic**- normal MCV- usually anemia of renal failure but sometimes represents iron deficiency anemia
 - Macrocytic**- high MCV- nutritional deficiencies (folate or B12), alcoholism, liver disease, or hypothyroidism

Incidental anemia without obvious source of bleeding or symptoms

- Anemia that is not severe (9 or more) found incidentally during an ED workup
- Arrange for outpatient followup
 - New anemia in an older patient is colon cancer until proven otherwise
 - Print a copy of the patient's lab work to bring to their followup appointment
 - Talk with the patient about the importance of following this up
 - Put a diagnosis of anemia on the chart and in your discharge instructions

Starting iron supplementation

- Younger females with heavy periods and microcytic anemia can be started on Iron supplementation if primary care or OB/GYN care will be delayed
- Start over the counter Ferrous Sulfate- 325mg once a day, gradually increase up to three times per day
- Best absorption on an empty stomach but this may cause nausea, vomiting or GI upset- usually best mitigated by slowly increasing dose
- Increased absorption with 250mg Vitamin C tablet or ½ glass of orange juice

Transfusing anemic patients

- Unstable patients with active bleeding- transfuse even with a normal or slightly low hemoglobin level
 - May take up to 24 hours for hemoglobin levels to drop after acute bleeding
 - Most concerning is patient who drops their hemoglobin over a span of several hours
 - Consider emergency release type O blood for unstable patients
 - Can use rapid transfuser to speed rate of transfusion and warm the blood
 - Verbally consent the patient for emergency transfusion- risks include infections such as HIV or Hep C (1 in a million), transfusion reaction, volume overload or fever (more common)
- Stable patients with hemoglobin levels at or near 7
 - Use a hemoglobin threshold of 7 for transfusion in nearly all patients
 - Multiple studies in different populations have shown increased morbidity and sometimes mortality with transfusion threshold of 9
 - Consider transfusion at 9 for those with active myocardial ischemia or infarction or those with severe and intolerable symptoms (chest pain, SOB)
- Logistics of transfusion
 - Send a type and screen to get a blood type/Rh and screen for rare antibodies
 - Type and crossmatch for anticipated number of blood units
 - One unit of pRBCs will raise hemoglobin by approx 1 point
 - Perform a written informed consent for the transfusion
 - Do this in the ED if possible to help your inpatient consultants
 - Don't necessarily need to start transfusion in the ED or do it quickly
 - Most of these patients bled down slowly so it's best to replace it slowly to avoid volume overload and other complications

PEARL: In general, a type and cross reserves those units for that particular patient. If you do not use the blood units within 72 hours, those units may be discarded so call your blood bank and cancel the cross-match if you don't actually use that blood to save this limited resource

Other anemia labs- can order these in the ED to help your admitting team- make sure they followup the results

- Iron panel (serum iron level, TIBC, transferrin, ferritin)
- Retic count
- Peripheral blood smear
- Direct and indirect coombs
- LDH and haptoglobin (for hemolytic anemia)

PEARL: In the ED, identification of the exact cause of anemia is usually unnecessary in stable patients who are not obviously bleeding (except for TTP- discussed later)

Hemophilia in the ED

- Most patients will already have a diagnosis but some may not
 - Suspect hemophilia in children with a family history of bleeding disorders or those patients with spontaneous or excessive bleeding out of proportion to the traumatic mechanism
- Hemophilia A- deficiency of Factor 8
- Hemophilia B- deficiency of Factor 9
- X linked inheritance so vast majority of patients are male, females are asymptomatic carriers
- Spectrum of disease- some patients have <1% factor activity, some have mild hemophilia with 30-40% factor activity

Labs

- PT normal, aPTT prolonged- except if mild hemophilia

Sources of bleeding

-At risk for bleeding anywhere (including into joints from sprains and fractures) but highest mortality across all ages groups is from intracranial bleeding

Imaging- have an extremely low threshold for aggressive imaging

- Non-contrast Head CT for all head trauma or headache- even minor or trivial mechanism
- CT abdomen/pelvis for any abdominal pain
 - At risk for retroperitoneal or iliopsoas hemorrhage
- MRI for any back pain- looking for epidural hematoma

Factor replacement

- Except for superficial abrasions or lacerations (even deep lacerations) you will need to give the patient factor replacement
 - Abrasions or lacs can be treated with direct pressure and topical thrombin
- Most patients should be replaced to 50% factor activity
 - Except GI bleeding and intracranial bleeding- replace to 100% factor activity
- Formula for replacing factors- patients should know baseline factor activity

-Factor 8- (Target activity level – baseline activity level) / (2 x pt weight in kilograms)

-Factor 9- (Target activity level – baseline activity level)/ (pt weight in kilograms)

Disposition- patients with minor bleeding into joints or soft tissue, oral/nasal mucos can be discharged after factor replacement, admit all others for continued monitoring and factor replacement

vonWillebrand's disease

- vonWillebrand's factor (vWF) is necessary for platelet aggregation
 - Most patients have decreased level of vWF, some have abnormal function of vWF, rare cases have virtually zero circulating vWF
- Most bleeding is epistaxis or gingival bleeding. Can also have unusual bruising, GI bleeding, or menorrhagia in menstruating females

Treatment

- Intranasal desmopressin for most cases
 - Children- 1 spray in either nostril
 - Adults- 1 spray in both nostrils
 - Fluid restrict for 24 hours- can develop hyponatremia
- Severe cases- cryoprecipitate or Factor 8 (both contain vWF)
 - Controversy as to best treatment so consult heme/onc for preferred treatment

Idiopathic Thrombocytopenic Purpura (ITP)

- Causes rapid destruction of platelets
- ITP is a result of auto-immune diseases, infections or certain medications
- Petechiae or purpura
 - Non-blanching purplish rash
 - Petechiae are 1-2mm in size and punctate
 - Purpura are larger splotches
- Patients usually notice the rash first but they may also have minor bleeding of the gingiva or menorrhagia

Labs- CBC should be normal except the platelet count- may have minor anemia but RBC indices should be normal

Treatment- depends on level of thrombocytopenia

- Greater than 50,000 platelet count (30K in kids)- no specific treatment required, ok for outpatient follow-up
- Less than 50,000 platelet count with active bleeding- admission for treatment or workup
- Less than 30,000 platelet count- admission for treatment or workup

Medications that can cause ITP

- Heparin, sulfa drugs, and chronic ethanol use are most common med cause of ITP
- Aspirin, rifampin, and penicillins have a less strong association with ITP

Treatment of ITP

- Steroids- Prednisone 60mg PO for adults
- Severe bleeding or platelet count less than 10,000- consider IVIG- 1 gram per kilogram per day
- In general don't transfuse platelets because ITP will just chew them up
- Only transfuse platelets for patients with life threatening bleeding
 - Be sure to give steroids first to decrease platelet destruction
 - Some recommend up to 1 gram of IV methylprednisolone, talk with your heme/onc consultant

Thrombotic Thrombocytopenic Purpura (TTP)

- Any patient with microangiopathic hemolytic anemia (MAHA) plus thrombocytopenia is TTP until proven otherwise
 - MAHA is diagnosed by seeing red blood cell fragments or schisocytes on a peripheral blood smear
- Simplified explanation of TTP
 - Dysfunctional protein called ADAMTS-13
 - Usually mops up circulating vWF
 - When it doesn't work- forms diffuse microclots

Diagnostic features

- FAT RN (no offense- it's really the best mnemonic!)
- Fever Anemia Thrombocytopenia Renal Failure Neuro symptoms

PEARL: Patients may not have all of these criteria or have them at the same time. DO NOT let the absence of any of these criteria to stop an appropriate workup for TTP. Untreated mortality for TTP is 80%

Causes- infection, auto-immune diseases, medications, and pregnancy

Treatment- plasmaphoresis- exchange patient's entire plasma volume or more

- Avoid platelet transfusions except in cases of life threatening bleeding or intracranial bleeding
- Can consider fresh frozen plasma (FFP) to temporize if there will be a delay in plasmaphoresis

PEARL: In any patient with thrombocytopenia and anemia, be sure to check a peripheral blood smear to check for schisocytes that's needed to diagnose TTP

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