



05/21/21 Morning Report with @CPSolvers



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<p>CC: Yellowish discoloration of eyes & fatigue</p> <p>HPI: 16 M p/w 1 month of fatigue & reduced appetite. He reports a yellowish discoloration of his eyes & skin and dark wine discoloration of urine.</p> <p>No wt loss, itching, drug intake, blood transfusion, heavy metal exposure</p>	<p>Vitals: T: 97F HR: 82 BP: 118/78 RR: 19 SpO₂: 98% RA</p> <p>Exam:</p> <p>Gen:</p> <p>HEENT: Pallor, icteric conjunctiva</p> <p>CV: S1, S2 + No rubs/murmurs/gallops</p> <p>Pulm: Vesicular breath sounds in all lung fields; No added sounds</p> <p>Abd: Mild Hepatosplenomegaly; No stigmata of CLD</p> <p>Neuro: No FND</p> <p>Extremities/Skin: Yellowish discoloration</p>	<p>Problem Representation: 16 yr M w/ no PMH presents w/ subacute fatigue, jaundice, dark urine, found to have hepatosplenomegaly on exams and labs suggestive of hemolytic anemia.</p>
<p>PMH: No similar episodes in the past</p> <p>Meds: None</p> <p>Fam Hx: Nothing significant</p> <p>Soc Hx: Nothing significant</p> <p>Health-Related Behaviors: No tobacco, alcohol intake</p> <p>Allergies: No known allergies</p>	<p>Notable Labs & Imaging:</p> <p>Hematology: WBC: 5 Hgb: 6 Plt: 150,000 MCV 101 Retic production index > 2.5; Direct coombs -ve</p> <p>Chemistry: BUN:12 Cr: 0.72 glucose: 110 AST: 70 ALT: 40 Alk-P: T. Bili: 4.5 Ind Bili 3.8 Albumin: 4.3 Globulin 2; INR 1.2 LDH 2000 HBV, HCV -ve, ANA -ve, G6PD N; CD55, 59 N; Osm fragility test N; Peripheral Smear: No parasites</p> <p>Imaging: USG Abdomen: spleen 16cm, mod hepatic enlargement, N portal V diameter Liver Bx: Chronic hepatitis changes, preserved architecture Slit lamp: Kayser-Fleischer ring; S. Ceruloplasmin 8; 24 Cu 132/24 hrs Liver Bx: Elevated copper concentration</p> <p>Dx: Wilson's Disease</p>	<p>Teaching Points (Gurleen):</p> <ul style="list-style-type: none"> ● FATIGUE, REDUCED APPETITE: no specificity ● HYPERBILIRUBINEMIA: <u>pre-liver</u> (indirect bilirubin - inc synthesis, dec uptake/conjugation) vs. direct bili: <u>intra-liver</u> (bilirubin formation & excretion) or <u>post-liver</u> (obstruction of biliary system-stone, cancer, PSC) -<u>Indirect bili:</u> hemolysis → pigmentation of urine -<u>Direct bili</u> (soluble): dark urine ● SPLENOMEGALY: water (portal hypertension), molecules (amyloid, lipid storage-Gaucher), RBCs (sickle cell, AIHA), or WBCs (infection, autoimmune, myeloprolif, drugs, malignancy) ● HEPATOMEGALY VENN DIAGRAM: portal hypertension (would see evidence), WBCs, or molecules ● INDIRECT HYPERBILI: inc delivery (hemolysis, hematoma), dec uptake (congestive hepatopathy, portosystemic shunts), dec conjugation -Gilbert's, hyperthyroid, meds) ● ELEVATED LDH: cell lysis. <i>Layer with stability:</i> liver, kidney, muscle, or RBC. <i>Layer w/ anemia, elv retic index:</i> RBC ● HEMOLYTIC ANEMIA -<u>Environment</u>-schistocyte(MAHA-would see thrombocytopenia, trauma) -<u>Membrane</u>-spherocyte (autoimmune, hereditary spherocytosis -osmotic fragility test or E5MB, cirrhosis-Zieve's, PNH) -<u>Intrinsic</u> (infections, enzyme deficiency, hemoglobinopathy) ● OVERLAP OF HEMOLYSIS + LIVER DISEASE (liver disease can make detection of hemolysis challenging): cirrhosis, Zieve's, Wilson's ● SMEAR NEG HEMOLYSIS: environment (smear + in only 50% in DIC, MacroAHA), cells (G6PD, electrolytes: hypophos, hyperCopper-Wilson)