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Immune System Overdrive: A Case Study of an Obstetric Patient with Hemophagocytic Lymphohistiocytosis

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This offering details the presentation, differential diagnosis and evidence-based interprofessional management of an obstetric patient afflicted with Hemophagocytic Lymphohistiocytosis (HLH).

CASE PRESENTATION
- 33 year old G2P1, 27 0/7 weeks
- Uncomplicated pregnancy; normal fetal growth ultrasound
- 8/21/16 hospital day (HD) #1 – transferred from a community hospital; admitted to a high-risk antepartum unit in a 1,200 bed Magnet® institution
- Symptoms – 6 day history of headache, fever, malaise, intermittent epigastric pain, and decreased appetite
- Sick contact – 2 year old daughter with febrile illness 1 week prior to admission
- Attributed on admission, stable BP
- Lab Abnormities – Platelets 94,000 (150,000-400,000), total bilirubin 3.1 (0.1-1.0), direct bilirubin 2.5 (0-0.2), AST 188 (<41), ALT 166 (<56), LD 623 (100-250), ferritin level 10,226 (10-291), triglycerides 1,981 (<150), creatinine 0.9 (0.6-1.5), hyponatremia (serum sodium 124), periodontal disease
- Fetal bradycardia
- Fever, neurologic abnormalities, cytopenias, hyperferritinemia, hepatosplenomegaly, hypertriglyceridemia, elevated liver enzymes, hemophagocytosis (bone marrow aspirate)
- Diagnostic test: hyperferritinemia associated with Hemophagocytic Lymphohistiocytosis
- Hospital day 2 – 8/22/16
- Fever to 103.0 at 0140, leukopenia with bandemia, maternal hypertension (HR 130–140) and altered mental status
- Transferred to ICU 8/22/16 at 0515
- Oxygen desaturation to 86–88%; addition of nitric acid therapy and high frequency ventilatory support – not tolerated, maternal hypotension, maternal (HR 130–140) and fetal (HR 170s) tachycardia, tachypnea, hypoxemia
- Interventions: ECMO
- Hospital Day 3 – 8/23/16
- New onset of encephalopathy at 1950, hypotension, hypoxemia, agitated
- Interventions: sedated, placed in a medically induced coma, blood pressure support
- Hospital Day 4 – 8/24/16
- Further respiratory decompensation, oxygen demand of chest pain/pressure and new onset of encephalopathy; addition of nitric acid therapy and high frequency ventilatory support – not tolerated, maternal hypotension, tachycardia
- Interventions: decision made for stat cesarean section at the bedside at 27 3/7 weeks for a 1170 gram living male infant, apgars 3/5
- Complications
- DIC
- Post-op Course
- DIC
- ECON Decompression 9/11/16
- Dialysis
- Discharged to home with specialty follow-up 9/20/2016 (HD#31)
- Goal - to control an overactive immune system
- Eltopside – chemotherapy medication
- Steroid therapy – Dexamethasone IV

Differential Diagnosis
- Atypical Hemolysis, Elevated Liver Enzymes, and Low Platelet (HELLP) syndrome
- Viral infection
- Thoracic/thrombocytopenic purpura (TTP)
- Autoimmune disorder

WHAT IS HLH?
- Incidence – rare; few cases in pregnancy reported in the literature
- Pathophysiology – excessive activity of the immune system, overproduction of inflammatory cytokines
- Diagnostic Findings – fever, neurologic abnormalities, cytopenias, hyperferritinemia, hepatosplenomegaly, hypertriglyceridemia, elevated liver enzymes, hemophagocytosis (bone marrow aspirate)
- Prognosis – high mortality rate; early diagnosis and treatment is essential as initial symptoms are often non-specific

ANTEPARTUM HOSPITAL COURSE
- Hospital Day 2 – 8/22/16
- Fetal fibroblasts for TTP, leukemia with thrombocytopenia, maternal hypertension, and fetal tachycardia
- Ferritin level 10,226 (10-291), complete blood count and platelets count
- Interventions: Diuretics, fluid and electrolyte, additional labs, infectious disease consult, BP 92/50, oxygen therapy
- Hospital Day 3 – 8/23/16
- New onset of encephalopathy at 1950, hypotension, hypoxemia, agitation
- Interventions: sedated, placed in a medically induced coma, blood pressure support
- Hospital Day 4 – 8/24/16
- Further respiratory decompensation, oxygen demand of chest pain/pressure and new onset of encephalopathy; addition of nitric acid therapy and high frequency ventilatory support – not tolerated, maternal hypotension, tachycardia
- Interventions: decision made for stat cesarean section at the bedside at 27 3/7 weeks for a 1170 gram living male infant, apgars 3/5
- Intravenous fluids and antibiotics, additional labs, infectious disease consult

POST-OP COURSE
- Complicated by:
- Dialysis
- Discharged to home with specialty follow-up 9/20/2016 (HD#31)

UNCOVERING THE DIAGNOSIS
- 8/25/16
- Ferritin level 10,226 (10-291)
- Triglycerides 1,981 (<150)
- 8/25/16
- Bone marrow biopsy – phagocytes suggestive of HLH

TREATMENT
- Goal - to control an overactive immune system
- Eltopside – chemotherapy medication
- Steroid therapy – Dexamethasone IV

LESSONS LEARNED
- Seek interdisciplinary support and management in order to provide high quality care to the critically ill obstetric patient.
- Recognize the need for a higher level of care and coordinate timely transport to a tertiary care facility to ensure optimal patient outcomes.
- Think ‘outside the box’ with pregnant patients – use your interprofessional resources to find answers.

REFERENCES