

HELLP Syndrome

Risk

- Occurs in 0.1–0.9% of all pregnancies.
- Previous history of preeclampsia or HELLP syndrome is a risk factor for HELLP.
- Occurs in 10–20% of women suffering from preeclampsia with severe features.
- May be a form of preeclampsia. Preeclampsia occurs in 2–10% of pregnancies; it is more prevalent in women with diabetes, women who are obese, and older women.

Perioperative Risks

- High maternal and fetal morbidity and mortality
- Increased cesarean delivery rate, increased intraop hemorrhage

Worry About

- Can be confused with hepatitis, thrombotic thrombocytopenic purpura, gallbladder disease, viral illnesses, antiphospholipid syndrome, and acute fatty liver of pregnancy.

- Thrombocytopenia and coagulopathy increase risk of hematoma associated with neuraxial anesthetic.
- High risk of hemorrhagic complications; associated with placental abruption.
- Upper airway and laryngeal edema can lead to airway obstruction and difficult or failed intubation. Fluid management can be difficult; pulm edema may ensue.

Overview

- HELLP is an acronym for the findings that suggest hepatic involvement in preeclamptic pts: Hemolysis, Elevated Liver enzymes, Low Platelets. It typically presents between 28–36 wk gestation but has been reported to occur postpartum.
- Diagnostic criteria include hemolysis, defined by abnormal peripheral smear/microangiopathic hemolytic anemia and increased bilirubin levels; elevated liver enzymes; and thrombocytopenia.
- Failure to treat may lead to eclampsia or death due to hepatic hematoma or rupture.

- Up to 20% of pts with HELLP syndrome do not have antecedent Htn.

Etiology

- Poorly understood; may be severe form of preeclampsia resulting from abnormal prostaglandin control, intravascular platelet activation, and microvascular endothelial damage

Usual Treatment

- Definitive treatment is delivery as quickly as possible.
- After delivery, many experience full recovery and plt counts returning to normal within 1 wk.
- Recent evidence challenges the role of glucocorticoid therapy.
- Platelets, FFP, and cryoprecipitate administered as needed.
- Magnesium sulfate for CNS irritability and antihypertensives for Htn.

Assessment Points

System	Effect	Assessment by Hx	PE	Test
HEENT	Upper airway edema	Dyspnea, voice change	Poor visualization on airway exam	Mallampati assessment
CV	LV failure	Dyspnea, desaturation	Adventitious sounds, desat	CVP and/or LVEDP
RESP	Resp depression	Magnesium administration	Decreased reflexes	MgSO ₄ level
GI	Liver swelling Subcapsular hematoma	Epigastric pain N/V		Elevated AST, ALT, LDH >600 IU/L
HEME	Thrombocytopenia Hemolytic anemia	Bruising Pallor, jaundice	Bleeding (IV site oozing)	Platelet count <100,000 Bilirubin >1.2 mg/dL Peripheral smear
RENAL	Acute renal failure	Oliguria		Elevated uric acid, BUN, serum Cr
CNS	Eclampsia, cerebral edema	Seizures		

Key References: American College of Obstetricians and Gynecologists, Task Force on Hypertension in Pregnancy: Hypertension in pregnancy. Report of the American College of Obstetricians and Gynecologists' Task Force on Hypertension in Pregnancy, *Obstet Gynecol* 122(5):1122–1131, 2013; Fitzpatrick KE, Hinshaw K, Kurinczuk JJ, Knight M: Risk factors, management, and outcomes of hemolysis, elevated liver enzymes, and low platelets syndrome and elevated liver enzymes, low platelets syndrome, *Obstet Gynecol* 123(3):618–627, 2014.

Perioperative Implications

Preoperative Preparation

- Obtain CBC, PT, PTT, fibrinogen, ALT, AST, LDH, BUN, and Cr.

Monitoring

- Consider arterial line and baseline ABG.
- Consider CVP if oliguria persists despite fluid administration or CHE.

Airway

- Assess airway early and repeat airway exam periodically.

- Laryngeal edema may preclude normal tracheal intubation in the event of emergency C-section. Videolaryngoscopy should be considered for intubation.
- Difficult intubation equipment should be immediately available.
- Consider preemptive epidural or continuous spinal cath before platelet count drops.

Induction

- Control neuraxial anesthesia with incremental dosing of catheter, if not contraindicated. Spinal techniques can be safely used in severe preeclampsia without coagulopathy.

- If GA is required, the hypertensive surge associated with ET intubation can be reduced by pretreatment with magnesium, antihypertensives, and/or opioids.

Adjuvants

- If significant Htn, antihypertensive therapy prior to laryngeal intubation.
- If receiving magnesium sulfate and needs GA, small doses of neuromuscular blocking agents with close neuromuscular blockade monitoring.

Hemochromatosis

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Risk

- Incidence of primary (hereditary) hemochromatosis: In some Caucasian populations, 10% are heterozygous carriers and 0.25–1% homozygous.
- Age: Clinical manifestations typically occur after age 40 in men and later in women due to the protective effect of menses.

Perioperative Risks

- Infection due to accumulation of iron in immune cells
- Glycemic disturbances

- Bleeding risk from low levels of clotting factors or platelet dysfunction
- Decompensated heart failure and/or arrhythmias

Worry About

- Iron deposition in the liver, heart, and endocrine glands leading to dysfunction

Overview

- Primary (hereditary) hemochromatosis is transmitted by genes, and secondary hemochromatosis is acquired.
- HH is an autosomal recessive disorder (HFE gene) that results in excess iron absorption.

- Once excess iron is absorbed, humans have no way to increase excretion. Iron accumulates in organs and results in cell damage. Because 90% of excess iron is deposited in the liver, it is often most affected.
- HCC is one of the most serious complications from untreated HH, responsible for 45% of deaths in pts with HH. Presence of cirrhosis is the greatest prognostic indicator for increased mortality.

Diagnosis

- Diagnosis is made by looking for elevated serum ferritin (>200 ng/mL in women and >300 ng/mL in