CCM Tutorial: HT complications of Preg & OBS aspect of CPR in pregnancy (HELLP Syndrome)

HELLP Syndrome

- incidence approximately 0.6% of deliveries
- 3.1–12% of patients with severe PET
- 70–92% of HELLP occurs antepartum
- 8–30% of HELLP postpartum
- 9–11% of HELLP develop symptoms < 27 wks
- 25–80% at term
- Up to 15–20% of HELLP, no antecedent symptoms of PET
- HELLP rare in the first trimester, case report

HELLP Syndrome

- Epigastric pain, nausea, vomiting, malaise +/- other symptoms of severe PET
- Haemolysis
  - LDH > 600IU/L, haptoglobin <0.3 g/L
- Elevated liver enzyme
  - AST or ALT > 70IU/L
- Low platelet
  - < 150 , <100,

Table 2: Differential diagnosis of the HELLP syndrome.

1. Diseases related to pregnancy
   - Benign thrombocytopenia of pregnancy
   - Acute fatty liver of pregnancy (AFLP)

2. Infectious and inflammatory diseases, not specifically related to pregnancy:
   - Virus hepatitis
   - Cholangitis
   - Cholecystitis
   - Upper urinary tract infection
   - Gastritis
   - Gastric ulcer
   - Acute pancreatitis

3. Thrombocytopenia
   - Immunologic thrombocytopenia (ITP)
   - Folate deficiency
   - Systemic lupus erythematosus (SLE)
   - Antiphospholipid syndrome (APS)

4. Rare diseases that may mimic HELLP syndrome
   - Thrombotic thrombocytopenic purpura (TTP)
   - Haemolytic uremic syndrome (HUS)

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Table 5 Differential diagnosis of HELLP syndrome, TTP/HUS, and AFLP [4, 52, 54, 114, 133–138]

	HELLP syndrome	TTP/HUS	AFLP[a]
Time of onset	Most commonly presents in the late third trimester, but has been reported as early as the second trimester	Any time	Most commonly presents in the late third trimester, but has been reported as early as the second trimester[a]
Coagulation studies	Present[b]	No	Present[c]
Thrombocytopenia	Present	Present	Present
Hypoglycemia	No	No	Yes
Renal failure	+/–	+	+
Hemolysis	+	+	–
History of preeclampsia	++/–	–	+/–

[a] History of the infant with FAO enzyme deficiency is relatively common

[b] Coagulopathy occurs in HELLP syndrome when it is complicated by DIC syndrome (consumption coagulopathy)

[c] Most commonly coagulopathy is associated with liver failure

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Table 3: Complications reported in the HELLP syndrome

Maternal complications	Occurrence (%)
Eclampsia	4–9
Abruptio placentae	9–20
DIC	5–56[1]
Acute renal failure	7–36
Severe ascites	4–11
Cerebral oedema	1–8
Pulmonary oedema	3–10
Wound hematoma/infection[2]	7–14
Subcapsular liver hematoma	Between 0.9% and <2%
Liver rupture	>200 cases or about 1.8%
Hepatic infarction	>30 cases combined with APS
Recurrent thrombosis	Associated with prothrombin gene 20210a mutation
Retinal detachment	1
Cerebral infarction	Few case reports
Cerebral Haemorrhage	1.5–40[3]
Maternal death	1–25
Foetal/neonatal complications	
Perinatal death	7.4–34
IUGR	38–61
Preterm delivery[4]	70 (15% < 28 gestational weeks)
Neonatal thrombocytopenia [5]	15–50
RDS	5.7–40

Complications as severe PET

Complications as severe PET

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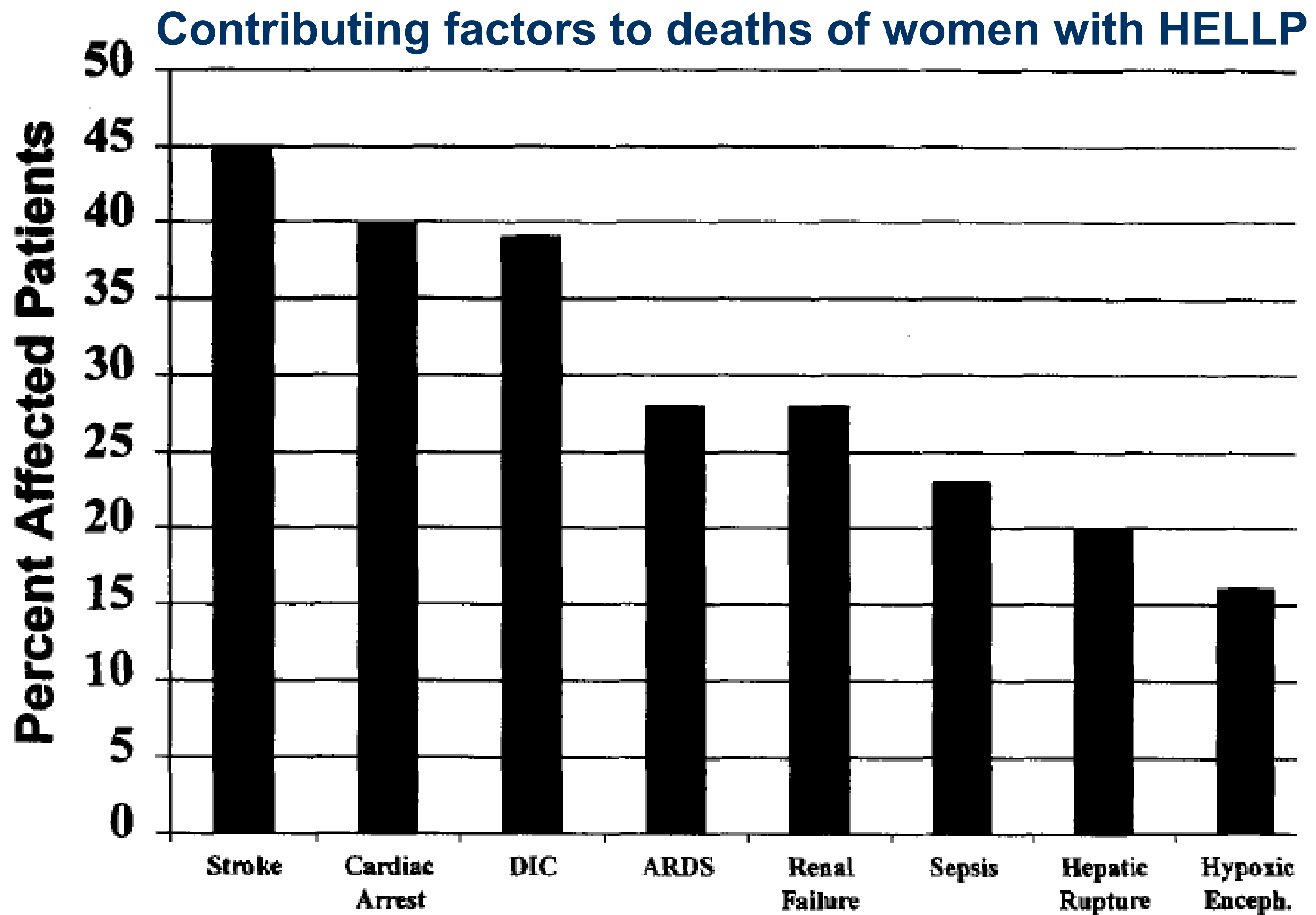

"Maternal mortality associated with HELLP syndrome". 54 Maternal death related to HELLP. Literature review. Am J Obstet Gyneco1999

Table 1: Main diagnostic criteria of the HELLP syndrome

HELLP class	Tennessee Classification	Mississippi classification
1	Platelets ≤ $100 \cdot 10^9$/L AST ≥ 70 IU/L LDH ≥ 600 IU/L	Platelets ≤ $50 \cdot 10^9$/L AST or ALT ≥ 70 IU/L LDH ≥ 600 IU/L
2		Platelets ≤ $100 \cdot 10^9$/L ≥ $50 \cdot 10^9$/L AST or ALT ≥ 70 IU/L LDH ≥ 600 IU/L
3		Platelets ≤ $150 \cdot 10^9$/L ≥ $100 \cdot 10^9$/L AST or ALT ≥ 40 IU/L LDH ≥ 600 IU/L

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The spectrum of severe preeclampsia: Comparative analysis by HELLP (hemolysis, elevated liver enzyme levels, and low platelet count) syndrome classification

Am J Obstet Gynecol 1999

- retrospective analytic study, 777 patients with class 1, 2, or 3 HELLP syndrome
- compared & contrasted with those of 193 women with severe PET but without HELLP syndrome.

- **CONCLUSION:** Laboratory and clinical indices of disease severity in patients with severe PET or eclampsia generally were highest with class 1 HELLP syndrome & lowest when HELLP syndrome was absent.

Management of HELLP

- Diagnosis
- Antenatal case: assessment of severity, need of antenatal corticosteroids for fetal lung, stabilization & +/- conservative management for 24-48 hours, delivery
- Peripartum management as severe PET
- Supportive treatment if needed
- ? High dose corticosteroids

Use of high dose steroids in HELLP

- Retrospective and small randomized studies
  - high-dose dexamethasone (10 mg dexamethasone Q12h) reduced maternal morbidity & induced more rapid improvement of the PLT counts
- RCT
  - 10mg dexamethasone Q12H, Prednisolone 50mg Q12h
  - did not reduce maternal complications: acute renal failure, pulmonary edema & oliguria
  - not reduce rates of platelets & FFP transfusions were not significantly reduced, nor was the
  - time of recovery of laboratory test shortened in selected studies

Conclusion

- HELLP is not common
- Delivery is the treatment of choice
- Supportive and intensive care
- High dose steroid: still uncertain to be benefit
- Appropriate management, maternal mortality is still low