

HELLP

Syndrome

- **H**aemolysis with microangiopathic blood smear
- **E**levated **L**iver enzymes
- **L**ow **P**latelet count

Incidence

- 0.1 to 0.2 % of pregnancies overall
- 10-20% of severe pre-eclampsia / eclampsia
- 7.4-34% mortality rate

Risk Factors

- Previous hx of HELLP or pre-eclampsia
- Genetic variants

Pathophysiology

- Immunological maladaptation likely triggers insult to invading trophoblast
- Anti-angiogenic factors released into maternal blood trigger vascular endothelium
- Inflammatory response -> generalised endothelial and microvascular injury
- Fibrin deposition, platelet aggregation. Fragmented red cells (intravascular haemolysis)

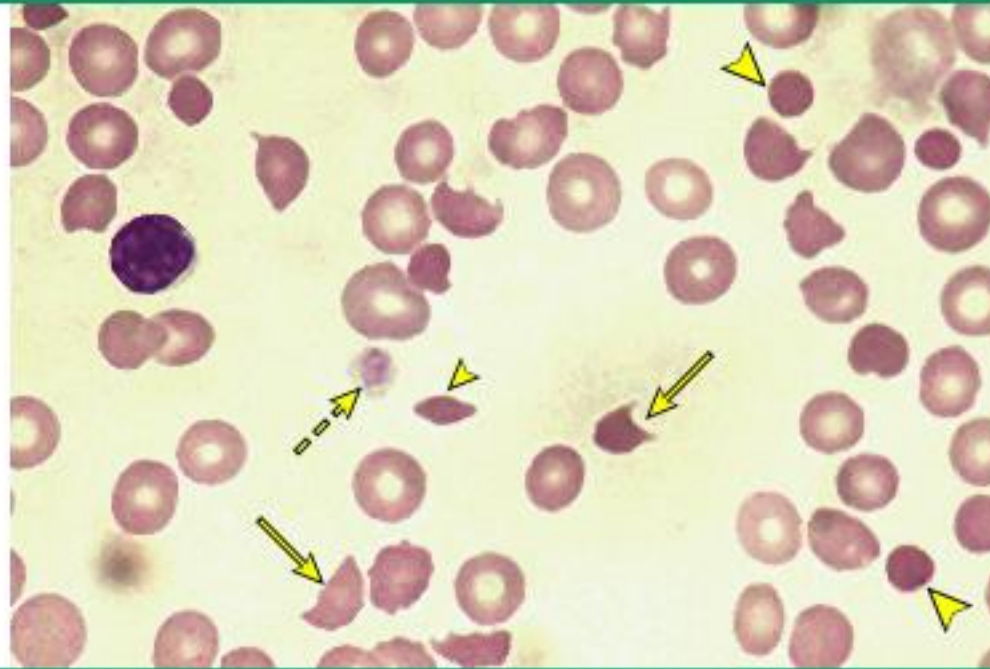
Presentation

- Rapid onset typically 28-36 weeks of gestation 10% before 27 weeks / 30% post-partum
- Excessive weight gain and generalised oedema
- Abdominal pain (tender epigastrium / RUQ)
- Nausea, vomiting, malaise
- Headache, visual changes, jaundice, ascites
- HTN and proteinuria may be absent

Tennessee classification

- Microangiopathic hemolytic anemia with schistocytes on blood smear.
- Platelet count $\leq 100,000$ cells/microL
- LDH > 600 IU/L
- Serum AST > 2 times upper limit of normal for local laboratory (or AST > 70 IU/L) .

Peripheral smear in microangiopathic hemolytic anemia showing presence of schistocytes



Peripheral blood smear from a patient with a microangiopathic hemolytic anemia with marked red cell fragmentation. The smear shows multiple helmet cells (arrows) and other fragmented red cells (small arrowhead); microspherocytes are also seen (large arrowheads). The platelet number is reduced; the large platelet in the center (dashed arrow) suggests that the thrombocytopenia is due to enhanced destruction.

Courtesy of Carola von Kapff, SH (ASCP).

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Table 1

Main diagnostic criteria of the HELLP syndrome

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

Complications

- Haemorrhage – placenta, PPH, liver haematoma, intracerebral / brainstem, DIC
- Infarction – liver, brain
- Pregnancy – fetal demise, pre-term delivery
- Other – visual impairment, pulmonary oedema, AKI

Management

Resuscitation

- Prepare for major bleeding: hepatic haemorrhage, subcapsular hematoma, liver rupture, MODS

Specific treatment

- Delivery if > 34 weeks or if baby and /or mother deteriorate
- Treat complications – APO, DIC, MODS
- Treat maternal HTN (labetalol / hydralazine / nifedipine)
- IV Magnesium for seizure prophylaxis
- IV steroids – given for fetal lung maturity from 24 to 34 weeks before delivery

- Liver haemorrhage – mx conservatively if possible, correct coagulopathy, surgical vs. arterial embolisation
- Plasma exchange – ongoing deterioration despite delivery
- Antithrombin and glutathione – no high quality trials yet
- Case reports of liver transplantation