



HELLP Syndrome vs Haemolytic Uraemic Syndrome in Pregnancy

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HELLP vs HUS

- Both HELLP syndrome and HUS present, haematologically, as a microangiopathic haemolytic picture.
- Both typically have falling/low platelets, high LDH and red cell fragments. Red cells are sheared by fibrin strands in the microvasculature. Appear as triangle and helmet form fragments.
- Onset, clinically, can vary in severity and rapidity, depending on many factors such as blood pressure (HELLP), dose of toxin (HUS).
- Both need to be monitored to avoid/treat DIC.

Incidence of HELLP and HUS in Pregnancy

- HELLP 0.2 – 0.6 %
- HUS 0.004 %

HELLP Syndrome

- Classic signs of **HELLP** syndrome are falling platelets, raised LDH and liver enzymes, and red cell fragmentaton. (*H*aemolysis, *E*levated *L*iver enzymes, *L*ow *P*latelets). Red cell fragments are of helmet and triangle forms (schistocytes).
- A severe condition of uncertain aetiology occurring in 0.2-0.6% of pregnancies.
- HELLP is generally listed with the pregnancy hypertension disorders, but relationship is uncertain. Although HT, PE and eclampsia do play a role in severity of condition and onset.

HELLP Syndrome

The pregnancy hypertensive disorders.

- **PIH** : pregnancy induced hypertension – persisting high blood pressure (150/100 to 180/110 mmHg), no proteinuria.
- **PE** : Pre-eclampsia – HT plus proteinuria, +/- oedema. If severe, then headaches, visual disturbances can occur.
- **Eclampsia** : HT, proteinuria, oedema, visual disturbances, seizures.

HELLP Syndrome

Recent studies and reviews have suggested that endothelial damage is the cause of **pre-eclampsia**, and that various organs can be affected.

- Endothelial damage within the kidney results in fenestration damage, leading to protein loss in the glomerulus.
- If the liver is affected, then patient will develop **HELLP** syndrome.
- Endothelial damage in the brain may lead to neurological sequelae such as seizures.

HELLP Syndrome

Assessment of Risk Factors

- A severe coagulopathy can result, so an extended coagulation screen, including PT, APTT, Fibrinogen and D-dimers (or equivalent) should be performed to assess status.
- LDH, Platelet count and coag profile need to be monitored.
- (Assessment of foetal lung maturity may be worthwhile, such as amniotic fluid surfactant/albumin ratio.)

HELLP Syndrome

Risk Reduction

- The most effective measure to reduce both maternal and foetal risk has always been to deliver the baby – in fact it is the delivery of the placenta which is responsible for the risk reduction.
- Corticosteroids may be given to help infant lung maturity, generally the day before delivery – particularly if gestation is less than 36 weeks.

Resolution is usually spontaneous, and occurs over a few days.

HUS

- Associated with a Shiga-like toxin produced by specific strains of E coli bacteria (eg O157:H7).
- Damage tends to focus on the kidney – namely platelet fibrin thrombi in the glomeruli and renal microvasculature, although uncommonly, it can be systemic.
- A severe coagulopathy can result, so an extended coagulation screen, including PT, APTT, Fibrinogen and D-dimers (or equivalent) should be performed to assess status.

HUS

Treatment

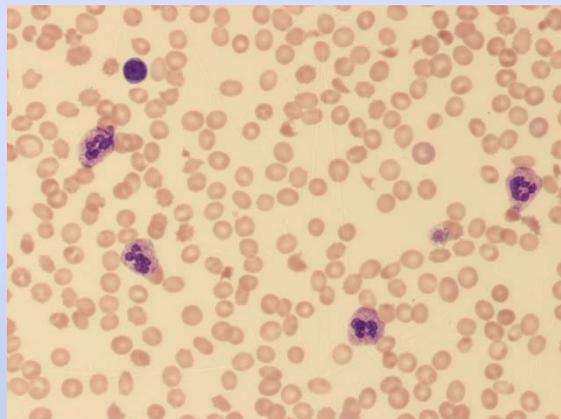
- Baby may be induced, to allow for less restricted treatment of HUS.
- Monitoring of coagulation profiles (D-dimers)
- Plasma exchange, corticosteroids, dialysis, platelet transfusions, FFP, clotting factors (all at need).
- Renal damage can be severe and permanent, with prompt treatment reducing morbidity.

(We are still treating patients affected by the Garibaldi-induced HUS out break 18 years ago.)

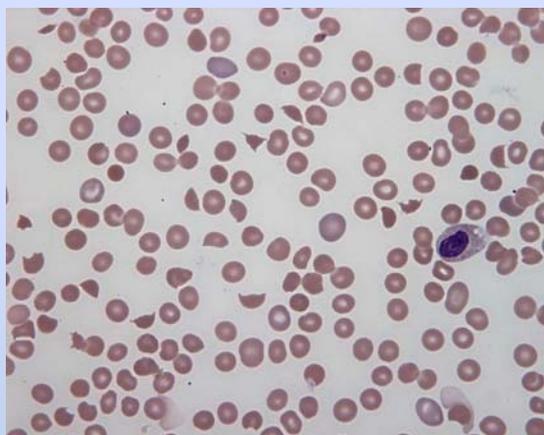
Case Studies

- HELLP – A 32 year old female, admitted to hospital at 36 weeks gestation with hypertension and suspected pre-eclampsia.
- HUS – 30 year old admitted to hospital at 36 weeks gestation with stomach cramps, diarrhoea. No pre-eclampsia. Had been to The Royal Adelaide Show.

HELLP Syndrome



HUS



HELLP Syndrome vs HUS

	HELLP Syndrome			HUS		
	Day 1	Day 2	Day 3	Day 1	Day 3 (am)	Day 3 (pm)
Sodium (131 – 142 mmol/L)	136	135	135	131	130	129
Potassium (3.3 – 4.7 mmol/L)	3.7	4.0	4.6	4.1	4.1	4.5
Chloride (97 – 109 mmol/L)	105	105	107	101	101	97
Bicarb (20 – 29 mmol/L)	18.2	19.7	22.0	18.4	18.4	24.7
Urea (1.2 – 4.0 mmol/L)	2.7	4.1	5.9	4.4	9.6	10.1
Urate (0.12 – 0.35mmol/L)	0.33	0.40	0.37		0.54	0.53
Creat (38 – 67 µmol /L)	68	87	99	65	255	239
Magnesium (Ther 1.7-3.5 mmol/L)	-	-	3.68			
Total Protein (58– 72 g/L)	61	63	50	61	50	45
Albumin (30 – 40 g/L)	31	31	25	27	22	20
Total Bili (2 – 24 µmol/L)	7	23	14	3	4	4
LDH (120 - 280 IU/L)	484	1054	594	253	962	825
GGT (5 - 30 IU/L)	78	96	70	44	25	23
ALT (5-30 IU/L)	96	117	83	24	24	24
ALP (50-215 IU/L)	194	160	119	232	152	130
Haemoglobin (110 - 160 g/L)	93	86	81	127	84	69
Platelets (150 – 450 x 10 ⁹ /L)	160	82	78	186	49	44
D-Dimers (<0.5)		1.5	0.8		1.5	
Red cell fragments	+	++	+	-	++	++

HELLP vs HUS

- Biochemically, HELLP tends to show more abnormalities in the liver enzymes, whereas HUS affects the kidneys, elevating Urea and Creatinine.
- The haematological pictures tend to look similar, although HELLP tends to have less red cell fragments.
- Clinical history is important, as HUS is caused by a toxin, whereas HELLP is of less certain origin, although recent studies are suggesting an endothelial problem. It is exacerbated by hypertension and pre-eclampsia.

HELLP vs HUS

- It is important, in seeing red cell fragments in the film, to ascertain if the platelet count is accurate. Are there fibrin strands, platelet clumps etc?
- To any existing biochemistry, add an LDH, LFT, renal function tests etc, that may differentiate the various haemolytic anaemias.
- If you suggest the possibility of a HELLP syndrome or HUS, then a repeat should be requested, including extended coag screen.

Treatment

In each case, the baby was delivered on day 3.

- In the HELLP syndrome case, resolution occurred spontaneously.
- In the HUS case, it was to allow more flexibility of treatment and dialysis. Although dialysis is generally quite safe in pregnancy, the added strain of the HUS increased the risk. Long term renal damage was minimal.

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Thank You