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 fragmentation. (Hemolysis, Elevated Liver enzymes, Low Platelets). Red cell fragments are of helmet and triangle forms (schistocytes).
- A severe condition of uncertain etiology 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 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 (e.g., 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
### HELLP Syndrome vs HUS

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

HELPP 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.

References

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Camille E. Powe, Richard J. Levine and S. Ananth Karumanchi.

Thank You