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# Acute fatty liver of pregnancy guideline (GL780)

**Approval**

| Approval Group | Job Title, Chair of Committee | Date |
|---|---|---|
| Maternity & Children's Services Clinical Governance Committee | Chair, Maternity Clinical Governance Committee | 5th October 2018 |

**Change History**

| Version | Date | Author, job title | Reason |
|---|---|---|---|
| 1.0 | January 2011 | Abdul Wagley (Specialist Registrar), Jill Ablett (Consultant Obstetrician | Trust requirement |
| 2.0 | June 2013 | Abdul Wagley (Specialist Registrar), Jill Ablett (Consultant Obstetrician | Reviewed |
| 2.1 | February 2016 | Jill Ablett (Consultant Obstetrician) | Reviewed – no changes |
| 2.2 | Sept 2018 | Jill Ablett (Consultant Obstetrician), Anna Ashcroft (ST6 Registrar) | Reviewed – no changes |

***Overview:*** Acute fatty liver of pregnancy (AFLP) is a rare but potentially fatal condition for both mother and baby, as often the diagnosis is delayed. It occurs in the latter part of pregnancy and may be part of a spectrum of disorders related to pre-eclampsia. It can cause multiple organ failure. Early diagnosis and prompt delivery is the mainstay of treatment. Referral to a specialist Liver unit or Intensive care may be required.

Incidence 1:20 000 maternities (1)

## Diagnosis

The condition is closely related to and can be difficult to differentiate from HELLP syndrome. See Table 1 (2)

| Symptom | HELLP | AFLP |
|---|---|---|
| Epigastric Pain | + | + |
| Hypertension | ++ | + |
| Proteinuria | ++ | + |
| Elevated Transaminases | + | ++ |
| Hypoglycaemia | +/- | ++ |
| Hyperuricaemia | + | ++ |
| DIC | + | ++ |
| Thrombocytopaenia | ++ | +/- |
| Leucocytosis | + | ++ |
| USS/CT | Normal | Bright Liver/ Ascites |
| Multiple Pregnancy | | + |
| Primiparous | ++ | + |
| Male Fetus | 50% | 70% (M:F = 3:1) |

***Table 1***

Widely accepted diagnostic criteria do not exist for AFLP; a set of standard diagnostic criteria has been proposed, but not evaluated in a large series as yet (3). The paper published by the UKOSS group (4) has recommended using these criteria referred to as the Swansea Criteria. See Table 2

This condition occurs predominantly in the third trimester with 74% diagnosed antenatally and the remainder postnatally (1).

The women at greatest risk are the older, primiparous, low BMI, multiple pregnancies.

Twin gestation carries 14 fold increased risk for AFLP.

**Common complications** [5] include:

- Renal failure (60%),
- Infection (45%)
- Coagulopathy (30%),
- Fulminant hepatic failure
- Hypoglycaemia (53%),
- Gastrointestinal haemorrhage (33%),
- Severe postpartum haemorrhage
- Stillbirth

| **Swansea Criteria** [3] | |
|---|---|
| 6 or more in the absence of another explanation | |
| Clinical (84%) | Vomiting (60%) |
| | Abdominal Pain (56%) |
| | Polydipsia/Polyuria (12%) |
| | Encephalopathy (9%) |
| Biochemical - Hepatic | Bilirubin (100%) >14 umol |
| | AST/ALT (100%) >42 IU/l |
| | Ammonia (50%) >47 umol/l |
| Renal | Urate (88%) >340 umol/l |
| | Creatinine (58%) >150 umol/l |
| Endocrine | Glucose (78%) < 4 mmol/l |
| Haematological | Leucocytosis (98%) $>11 \times 10^9$/l |
| | Coagulopathy- PT >14 secs OR APTT > 34 secs (often with Plt count $>100 \times 10^{12}$) (>50%) |
| Radiological – Abdominal USS | Bright Liver echo texture/Ascites (25%) |
| Histological – Liver Biopsy | Microvesicular steatosis |

***Table 2***

*In brackets is the percentage of patients with the abnormality according to the UKOSS study.* [1]

**Management** [6]

- Supportive Care in a multidisciplinary team ( inform obstetric , anaesthetic and ITU consultants)
- Aggressive management of **coagulopathy** with FFP, Crypoprecipitate, Novoseven in discussion with haematologist
- Aggressive management of **hypoglycaemia** with 50% intravenous glucose
- Expedite **delivery** if antenatal
- Mode of delivery
  - UK practice favours Caesarean section (perceived severity of condition, coagulopathy)
- Mode of anaesthesia

  - GA – negative effect on hepatic encephalopathy
  - Regional – risk of spinal haematoma in presence of coagulopathy
- Consider N-acetyl Cysteine
  Work extrapolated from acetaminophen-related hepatoxicity shows that N-A C improves haemodynamics whilst preventing progressive decompensation.
- Infection risk, hence low threshold for parenteral antibiotics.
- Reduce cerebral oedema by:
  - Raising the head of the bed, use of IV sedation, oral Lactulose, Hyperventilation.

**Information for counselling**

| Maternal Case fatality rate | 1.8% |
|---|---|
| Severe maternal morbidity | 28% |
| ICU admission | 60% |
| Specialist Liver Unit Admission | 18% |
| Mean Duration of stay | 9 days |
| Recurrence rate very low, linked to carriers of ß fatty acid oxidation genetic abnormality (LCHAD) | |
| Perinatal mortality rate | 104/1000 births, 10x national average |
| Stillbirth rate | 9% |
| Neonatal Case Fatality rate | 2% |

**References**

1. M. Knight, C Nelson-Piercy, J J Kurinczuk et al. A prospective national study of acute fatty liver of pregnancy in the UK. Gut 2008;57:951-956.
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3. C L Ch'ng, M Morgan, I Hainsworth, et al. Prospective study of liver dysfunction in pregnancy in Southwest Wales. Gut 2002;51:876-880.
4. https://www.npeu.ox.ac.uk/ukoss/completed-surveillance
5. G Yucesoy, S O Ozkan, H Bodur et al. Acute fatty liver of pregnancy complicated with disseminated intravascular coagulation and haemorrhage: a case report. International Journal of Clinical Practice 2005. Supplement (147):82-4.
6. J F Trotter. Practical management of acute liver failure in the intensive care unit. Current Opinion in Critical Care 2009;15:163-167.