## 1. Introduction and Who Guideline applies to

Acute fatty liver of pregnancy (AFLP) is a rare (1 in 20,000 maternities) but potentially fatal condition for both mother and baby, as often the diagnosis is delayed. It occurs predominately in the third trimester with 74% diagnosed antenatally and the remainder postnatally (1) and may be part of a spectrum of disorders related to pre eclampsia. It can cause multiple organ failure. Common complications include renal failure (60%), infection (45%), Coagulopathy (30%). Fulminant hepatic failure, hypoglycaemia (53%), GI haemorrhage (33%), Severe PPH and stillbirth. (2) Early diagnosis, stabilisation and prompt delivery is crucial.

The women at greatest risk are those who are older, primiparous, have a low BMI or those with multiple pregnancies. Twin pregnancy carries a 14 fold increase risk of AFLP.

## Scope:

This guideline is intended for use by medical staff, midwives and other relevant health care professionals. The severe consequences of AFLP are well established and obstetricians and other health professionals caring for pregnant women need to be aware of early diagnosis and interventions that may reduce the risk of morbidity and mortality.

## 2. Guideline Standards and Procedures

### Recommendations:

1. Diagnosis should be made as early as possible
2. Management should be aggressive
3. Delivery should be expedited if AFLP is diagnosed in the antenatal period

### Recommendation One:

Diagnosis should be made as early as possible

- The condition is closely related to and can be difficult to differentiate from HELLP Syndrome. (see table 1 below)
- Widely accepted diagnosis criteria do not exist for AFLP. The paper published by the UKOSS group (4) has recommended using the Swansea criteria (3) in table 2.
- Ultrasound should be performed to exclude other causes of jaundice such as hepatic infarct / haematoma.

**Table 1**

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

**Table 2**

| Swansea Criteria | 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%) >150umol/l |
| Endocrine | Glucose (78%) <4 mmol/l |
| Haematological | Leucocytosis (98%) > 11 x $10^9$ / l |
| | Coagulopathy – PT >14 secs **OR** APPT > 34 secs (often with Pl count >100 x $10^{12}$) (>50%) |
| Radiological – Abdominal USS | Bright liver echo texture / Ascites (25%) |
| Histological – Liver biopsy | Micro vesicular steatosis |

**Table 2 –** In brackets is the percentage of patients with the abnormality according to the UKOSS study

**Recommendation Two:**

Management should be aggressive

- The management should be by a multi-disciplinary team. (i.e. Obstetrician, Anaesthetist and the ITU Consultant)
- There should be strict fluid balance management in order to reduce pulmonary oedema
- As there is a high risk of developing coagulopathy and bleeding complications there should be early discussion with the Haematologists and precautions should be taken to prevent PPH. The management of coagulopathy should be with FFP, Cryproprecipitate,

Novoseven and this should be in discussion with the haematologist.

- Hypoglycaemia should be managed with 50% IV glucose until the LFT's are normal
- Consider N-acetyl Cysteine to improve haemodynamics whilst preventing progressive decompensation
- There should be a low threshold for parenteral antibiotics
- Cerebral oedema should be reduced by raising the head of the bed, use of IV sedation, oral lactulose and hyperventilation.

**Recommendation Three:**

Delivery should be expedited if AFLP diagnosed in the antenatal period

- The decision regarding timing and mode of delivery should be made with the MDT (Intensivists, Anaesthetists and Maternal Medicine / Obstetric teams)
- Mode of delivery should be by Caesarean section
- Mode of anaesthesia should be considered:
  - GA – negative effect on hepatic encephalopathy
  - Regional – risks of spinal haematoma in presence of coagulopathy

**Recommendation Four**

Need for continuous monitoring following delivery due to the risk of transient worsening of liver and renal functions and coagulopathy peripartum (5)

**Postpartum Management:**

In most patients, AFLP usually resolves completely after delivery, with return of normal liver function within 7 to 10 days [5] but the risk of transient worsening of Liver function tests remains. Liver function and coagulopathy typically begin to improve within days after delivery.

- Liver function tests, creatinine, and coagulation tests should be carried out every six hours until downward trend is observed, after which the frequency of testing can be reduced
- Some patients have a prolonged course with multi-organ failure, requiring supportive management in an intensive care unit, including mechanical ventilation, dialysis for acute renal failure, nutritional support because of associated pancreatitis, or transfusion of blood products for ongoing hemolysis or postpartum hemorrhage from atony or incisional bleeding.
- Management is carried out with the involvement of MDT( Anaesthetists, Maternal Medicine / Obstetric teams, Intensivists, Haematologists)

**Information for counselling**

Acute fatty liver can recur in subsequent pregnancies, even if testing for LCHAD mutation is negative. Women with a history of AFLP who are contemplating another pregnancy should be co-managed with a maternal-fetal medicine specialist.

| | |
|---|---|
| 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, 10 x national average |
| Stillbirth rate | 9% |
| Neonatal Case Fatality rate | 2% |

**Screening the infant for the gene defect should be discussed with the Neonatologists as this can be lifesaving if the mother is a carrier for the gene mutilation.**

## 3. Education and Training

None

## 4. Monitoring Compliance

| What will be measured to monitor compliance | How will compliance be monitored | Monitoring Lead | Frequency | Reporting arrangements |
|---|---|---|---|---|
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## 5. Supporting References (maximum of 3)


1. M Knight, C Nelson-Piercy, J J Kurinczuk et al. A prospective national study of acute fattl liver of pregnancy in the UK. Gut 2008:57:951-956.

2. 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): 82-84.

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. Nelson DB, Yost NP, Cunningham FG. Acute fatty liver of pregnancy: clinical outcomes and expected duration of recovery. Am J Obstet Gynecol 2013; 209:456.e1.


## 6. Key Words

Fatty liver, pregnancy, acute fatty liver

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Guidelines issued and approved by the Trust are considered to represent best practice. Staff may only exceptionally depart from any relevant Trust guidelines providing always that such departure is confined to the specific needs of individual circumstances. In healthcare delivery such departure shall only be undertaken where, in the judgement of the responsible health professional' it is fully appropriate and justifiable – such decision to be fully recorded in the patient's notes.

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**As part of its development, this policy and its impact on equality have been reviewed and no detriment was identified.**

| DEVELOPMENT AND APPROVAL RECORD FOR THIS DOCUMENT | | |
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| **Author / Lead Officer:** | F Siddiqui and L Matthews | **Job Title:** Consultant Obstetrician and Clinical Risk and Quality Standards Midwife |
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| **Approved by:** | Guidelines Group and Maternity Service Governance Group | **Date Approved:** 1.06.15 and 12.07.15<br>6.03.17 and 15.03.17 |

| REVIEW RECORD | | | |
|---|---|---|---|
| **Date** | **Issue Number** | **Reviewed By** | **Description Of Changes (If Any)** |
| January 2017 | V1 | F Siddiqui | No change |
| September 2020 | V2 | F Siddiqui and F Shakeel | Recommendation four added, postpartum management added. Women with a previous pregnancy with acute fatty liver should be managed by a maternal-fetal medicine specialist. |
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| DISTRIBUTION RECORD: | | | |
|---|---|---|---|
| **Date** | **Name** | **Dept** | **Received** |
| 7.15 | All Obstetricians and Midwives | Maternity | |
| March 2017 | All Obstetricians and Midwives | Maternity | |
| September 2020 | All Obstetricians and Midwives | Maternity | |
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