Diagnostic and Management Challenges of Acute Fatty Liver of Pregnancy: A Case Series

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Abstract

Introduction: Acute fatty liver of pregnancy remains an elusive and potentially fatal complication of a rare liver disease in late pregnancy. Due to sparse research, diagnostic dilemmas, and multi-organ involvement of the disease, it is imperative to elaborate the evaluation and management principles to confine the outcomes. **Case Reports:** Here, we aim to discuss in detail the peripartum manifestations and goal-directed management protocols in a case series of four patients managed at a tertiary care setup in India. **Conclusion:** It was found that all four cases had varied presentations ranging from asymptomatic hypoglycemia to liver failure and multiorgan dysfunction requiring a multidisciplinary perioperative care approach to prevent complications by incorporating supportive care along with latest available point of care diagnostics, interventional care, Extracorporeal Membrane Oxygenation (ECMO) and Continuous renal replacement therapy (CRRT).

Keywords: Acute fatty liver of pregnancy, cesarean section, liver diseases, pregnancy complications

Introduction

Acute fatty liver of pregnancy (AFLP) is an uncommon and potentially morbid obstetric emergency occurring secondary to microvesicular fatty infiltration of the liver leading to failure and multiorgan complications.[1]

It is a diagnostic dilemma due to vague presentations, relatively unknown pathophysiology, and similarity to various diseases such as hemolysis elevated liver enzymes low platelet (HELLP), intrahepatic cholestasis of pregnancy (IHCP), and preeclampsia. Although it has a prevalence of 1–3 cases per 10,000 deliveries, it has been shown to have high rates of fetomaternal complications and mortality (up to 18% and 26%, respectively).[1]

However, the development of multidisciplinary teams, early diagnosis employing Swansea criteria, and advanced critical care interventions can result in improved fetal and maternal outcomes.[1] In this case series, we aim to elaborate upon management and review of such cases at a tertiary care setup.

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Case Reports

The following cases of acute fatty liver are presented in detail after obtaining the Institutional review board approval vide CH (EC) Kolkata, letter no 8776/23 dated December 28, 2023. The individual consent has been obtained for the same.

Case 1 (A1)

A 24-year-old primigravida with diamniotic/dichorionic (DADC) twin pregnancy (post intrauterine insemination), hypothyroid on eltroxin, gestational hypertension (GHTN), and gestational diabetes mellitus (on medical nutrition therapy) presented at 35 weeks 5 days period of gestation (POG) with pain abdomen and jaundice for 1 week. On evaluation, the patient was found to have drowsiness, and icterus with bilateral pedal edema. Investigations revealed leukocytosis, thrombocytopenia with deranged coagulation profile (international normalized ratio [INR] – 2.91, D-dimer and fibrin degradation products [FDP]

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positive). Liver function test revealed, bilirubin of 14 mg/dl, serum glutamic oxaloacetic transaminase (SGOT);96U/L, serum glutamic pyruvate transaminase SGPT;151U/L, hypoalbuminemia, negative viral markers and mild azotemia (creatinine – 1.69 mg/dl). Ultrasonography (USG) revealed hyperechoic liver and diagnosed as a case of AFLP with coagulopathy. Emergency preanesthetic checkup (PAC) was done and the patient was accepted in the American Society of Anesthesiologists (ASA) Grade III (E) for emergency cesarean under general anesthesia (GA) with six fresh frozen plasma (FFP), four cryoprecipitates with steroid and appropriate blood demand before surgery. In OT, advanced hemodynamic monitoring (FloTrac, Edward Sciences) ensued, adequate venous access was secured and the patient was intubated using propofol and atracurium and maintained with desflurane. Intraoperatively, the patient was found to be hypertensive, and intraoperative blood loss was managed with blood products (2 packed red blood cells [PRBC], 2 FFP, 2 cryoprecipitates, and 4 random donor plasma [RDP]), tranexamic acid (TXA), and uterotonics, whereas delivered twins required minimal resuscitation. The patient was reversed and extubated, however, she required oxygen while being shifted to intensive care unit (ICU) for postoperative monitoring. In ICU, the patient continued to have tachycardia, pallor, hypertension (HTN), wound oozing with increased drain output, and intermittent hypoglycemia and managed with blood products (1 PRBC, 2 RDP and recombinant factor VII [rFVII] 4 units), high-flow nasal cannula (HFNC), labetalol, magnesium sulfate ($MgSO_4$), Vitamin K, dextrose, and antibiotics. She was later taken up for emergency laparotomy under GA in ASA IV (E) for hemoperitoneum, uncontrolled HTN, AFLP, and disseminated intravascular coagulation (DIC) under high-risk consent. The intraoperative period was uneventfully managed with hemodynamic monitoring and blood products (2 PRBC, 4 FFP, and 1 single donor platelet (SDP). Postoperatively, the patient developed hepatic encephalopathy (HE), hypostatic atelectasis, and leukocytosis managed with upgraded antibiotics, HFNC, albumin, electrolyte correction, and lactulose. The patient gradually recovered after conservative management and shifted to ward on postoperative day (POD)-11 with healthy babies.

Case 2 (A2)

A 43-year-old elderly primigravida, post- In vitro fertilization (IVF) DADC twin pregnancy (infertility and postcervical cerclage on ecosprin and progesterone), hypothyroid on eltroxin, presented at 33 weeks 6 days POG with jaundice, pain abdomen with constipation, and swelling of upper and lower limbs for 3 days. The patient was diagnosed as AFLP and admitted for early viable termination of pregnancy. On PAC evaluation, the patient was found to have peripheral edema and icterus. Investigations revealed raised bilirubin;14.6 mg/dl, SGOT;1220 U/L, SGPT; 1120 U/L, leukocytosis;14,300/mm^3, borderline coagulation profile with INR of 1.37 and negative viral markers, USG showed hyperechoic bright liver. The patient was taken up in ASA Grade III for elective cesarean section under GA following optimization with steroids, Vitamin-K, antibiotics, and lactulose. Appropriate blood demand was simultaneously placed. Intraoperatively, oozing appeared from the surgical field with the atonic uterus and blood loss (2 L) was managed with balanced crystalloids, TXA, uterotonics, and blood transfusion (2 PRBC and 4 FFP), however, bleeding could not be stopped and the patient underwent hysterectomy and shifted to ICU postextubation for further monitoring and management. In ICU patient was detected to have borderline HTN, sepsis (total leukocyte count [TLC] – 36,200/ mm3 and pro-calcitonin – 2.86 ng/ml) with coagulopathy (FDP and D-dimer positive) managed with upgraded antibiotics, laxatives, and blood products (2 FFP and Vitamin-K). General conditions improved, laboratory parameters stabilized and the patient was shifted on POD-3 to the ward.

Case 3 (A3)

A 22-year-old obese primigravida (postovulation induction and timed intercourse) with no known comorbidities presented at 36 weeks 5 days POG with complaints of jaundice, fever, headache, vomiting, and reduced appetite for 3–4 days duration. On evaluation, she was found to have a body mass index (BMI) of 34.3 kg/m^2, icterus, peripheral edema, and high blood pressure. 17,180/mm^3 with normal platelet and coagulation profile, direct hyperbilirubinemia (10.2 mg/dl) with transaminitis (SGOT/SGPT: 208/212 U/L), normal blood sugar, increased lactate dehydrogenase – 819 U/L, and bright liver on USG and negative viral markers and culture reports. The patient was diagnosed as a case of AFLP/severe preeclampsia with partial HELLP planned for early viable termination of pregnancy under steroid cover with induction of normal labor. The patient had failure of induction with nonprogress of labor and was taken up for emergency cesarean section. Standard GA protocol was followed, intraoperatively, the patient had HTN, oozing from the surgical site with a flabby uterus managed with antibiotic cover, intravenous (IV) fluids, $MgSO_4$ infusion, and uterotonics. The neonate delivered had a low APGAR score managed with positive pressure ventilation and was stabilized. The patient was extubated and shifted to ICU for postoperative monitoring and further management. In the initial postoperative period, blood pressure stabilized, however, anemia developed with the gradual increase in drain output with abdominal distension, generalized edema, raised leukocyte count, and gradual decrease in urine output with azotemia managed with central venous access, restricted IV fluids, diuretic trial, blood products (2 PRBCs), Vitamin-K, TXA, antibiotics (renal adjusted), and laxatives. By POD-3, the patient developed altered sensorium with intermittent hypoglycemia, sepsis (TLC – 29,460/mm^3) with coagulopathy (platelets – 60,000/mm^3, INR – 1.6, activated partial thromboplastin time – 57.5 seconds, FDP and D-dimer positive). It was accompanied by borderline hypotension and hypoxia with evidence of B/L basal synpneumonic effusion on lung USG and chest X-ray [CXR]). There was anuria with increasing azotemia, constipation and abdominal distension, patient was diagnosed as AFLP with DIC, sepsis and multi-organ dysfunction syndrome (MODS).

. The patient was managed with upgraded antibiotics, N-acetyl cysteine, dextrose, and albumin with frusemide infusion, blood products (4 FFP, 4 cryoprecipitates, and Vitamin K) intermittent hemodialysis with CytoSorb filter, laxatives, and HFNC/intermittent noninvasive ventilation support.

By POD-3, the evening condition worsened with hypotension, respiratory distress, anuria, bleeding diathesis, and severe metabolic acidosis. The patient was managed with elective mechanical ventilation, vasopressor support under continuous cardiac output (FloTrac, Edward Sciences) monitoring, steroid cover, broad-spectrum antibiotics, continuous renal replacement therapy (CRRT) with oxiris cytotoxin filter, and blood transfusion (2 PRBC, 6 FFP, 6 cryoprecipitates, and 1 SDP). Patient initially showed improvement in sensorium, blood parameters and in arterial blood gas (ABG) picture with tapered vasopressor support. On POD-05, patient started to develop acute respiratory distress syndrome (ARDS) with pleural effusion and respiratory acidosis. Patient was managed with pleurocentesis, lung protective ventilation followed by attempts of prone ventilation. However, due to hemodynamic instability (high vasopressor support), the patient suffered cardiac arrest and achieved the return of spontaneous circulation after 3 cycles of resuscitation. Due to poor lung and heart status, the patient was managed with extracorporeal membrane oxygenation (ECMO) therapy. On POD-6th after buildup with blood (4 PRBC and 1 SDP) and added inotrope support, the patient had an initial improvement in general condition and ventilation; however, due to severe disease progression (worsening ARDS, liver failure, precipitation of atrial fibrillation, multiple ecchymoses, and acute limb ischemia), despite all state-of-the-art intervention (ECMO, CRRT, multiple blood transfusion, antibiotics, etc.), the patient's condition deteriorated over the next few days and succumbed to her illness on POD-14.

Case 4 (A4)

A 22-year-old primigravida was admitted at 36 weeks 3 days POG with complaints of jaundice, vomiting, and high blood pressure for 3 days for early viable termination of pregnancy. Emergency PAC revealed no history of fever/localizing symptoms/focal neurological deficit/bleeding diathesis. The patient had borderline high blood pressure (150/88 mmHg) with normal airway and spine. Investigations revealed low platelets (80,000/mm^3) with borderline leukocytosis (11,500/mm^3) and coagulation profile (INR – 1.12), hyperbilirubinemia (Total serum bilirubin – 3.4 mg/dl) with mild transaminitis (SGOT/SGPT – 67/113 U/L), urine proteins were negative, ABG revealed compensated metabolic lactic acidosis (pH – 7.301, HCO_3 – 14.2, pCO_2 – 21, BE – −13.8, and lactates – 5.7), and asymptomatic hypoglycemia (random blood sugar [RBS] – 57mg/dl).

She was started on dextrose IV bolus (25%, 25 ml), administered antibiotics, antenatal steroids, and $MgSO_4$ infusion. The patient was diagnosed as AFLP/GHTN/HELLP syndrome and was accepted in ASA Grade III for emergency cesarean under neuraxial anesthesia. Subarachnoid block containing Inj bupivacaine heavy 0.5% 1.5ml and Inj fentanyl 25µg was administered with 27 G Quincke spinal needle. An adequate level of blockade was achieved; however, RBS was found to be 27mg/dl (asymptomatic) and continued to be in the hypoglycemic range despite 25% dextrose IV boluses. 10% dextrose fluid resuscitation commenced through central venous access. The surgery went uneventful with mild oozing from the surgical side controlled with TXA (1 g IV over 15 min) and pitocin infusion. A live healthy baby was delivered. Metabolic acidosis and hypoglycemia (RBS – 69 mg%) were corrected with 10% dextrose infusion and the patient was shifted to the ICU for postoperative monitoring and further management. In the ICU, the patient remained stable on dextrose-rich fluids and oral augmentation for borderline blood sugar levels. By POD-5, the patient was shifted to the ward.

The summary of the cases is provided in Table 1 and the snapshot of the Swansea criteria of the cases is given in Table 2.

Discussion

AFLP is a rare obstetric emergency that is potentially fatal when diagnosed late with inadequate management.[2] Data from publications in the 1980s suggested mortality rates in excess of 70%, but more recent estimates are dramatically lower with maternal mortality of 7%–18% and fetal mortality of 9%–23%.[3,4] The recognition of milder presentations, early intervention with delivery, and aggressive management of complications have likely contributed to a decreased mortality rate.[3,4]

The pathophysiological understanding remains elusive, involving both the mother and fetus.[1,5] The physiological changes during pregnancy are associated with a physiologic decrease in the oxidation of long- and medium-chain fatty acids, as well as fetal fatty acid oxidation disorders (most common-autosomal recessive deficiency of long-chain 3-hydroxyacyl-coenzyme A dehydrogenase) resulting in an increased maternal susceptibility to an overwhelming burden of free fatty acids leading to maternal hepatotoxicity and mitochondrial dysfunction over the course of gestation.[1,3-5] One of the hallmark findings in patients with AFLP is multiorgan fatty infiltration resulting in multiorgan failure and placental and endothelial dysfunction.[3]

Risk factors for AFLP include primigravida, multiple gestations, male fetuses, preeclampsia, high BMI, fatty acid oxidation disorders or growth retardation in the fetus, other liver diseases of pregnancy, and previous episodes of AFLP.[1,3,5-7] The potential factors influencing adverse maternal outcomes are male fetus, postpartum diagnosis of AFLP, intrauterine fetal death, DIC, younger mother (27 ± 2.57 years), singleton pregnancy, higher mean values of alanine transferase and bilirubin, and lower mean value of prothrombin activity.[1,7]

The presentation and laboratory findings of AFLP are vague, vivid, and nonspecific ranging from early mild asymptomatic cases to a severe overt hepatic failure with encephalopathy

posing a major diagnostic challenge.[2-4,6] The prodrome presents with generalized symptoms such as nausea/vomiting, abdominal pain, malaise, jaundice, anorexia, fatigue, and cold food preference in descending order in up to 70%–90% of patients.[4,7-10] It can progress to icterus, headache, polydipsia, pruritis, edema, ascites, encephalopathy, asterixis, HTN, and bleeding diatheses with concurrent HELLP syndrome or preeclampsia.[3,5] It usually occurs during the third trimester (30–38 weeks) to the postpartum period and seldom in the second trimester of pregnancy with average gestational age of 35.26 ± 2.58 weeks or 2.33 ± 0.57 days postpartum.[5,7-8,11]

The classic laboratory findings include deranged liver function tests with high bilirubin levels, deranged renal function tests, hemolysis, hyperuricemia, leukocytosis (neutrophilia) with

Table 1: Patient characteristics

Characteristic	Case A1	Case A2	Case A3	Case A4
Age (years)	24	43	22	22
BMI (kg/m^2)	29.1	29.6	34.3	21.2
Pregnancy	G1	G1	G1	G1
POG (weeks)	35.5	33.6	36.5	36.3
Gestation	Twin	Twin (post-IVF)	Singleton	Singleton
Comorbidities	Hypothyroidism, GHTN, and GDM	Infertility, hypothyroidism	Infertility	-
Previous AFLP	-	-	-	-
Presentation	Pain abdomen, jaundice (1 week)	Jaundice, pain abdomen and generalized edema (3 days)	Fever, vomiting, jaundice, osmotic (4 days)	Jaundice, vomiting, HTN (3 days)
Diagnosis	Swansea - 9/14	Swansea - 6/14	Swansea - 10/14	Swansea - 6/14
Obstetric Management	LSCS-same day, hysterectomy (POD-1)	LSCS and hysterectomy-next day	Trial of induction of labor followed by LSCS	LSCS
Anesthesia and ICU Management	GA, hemodynamic monitoring (CVC + IBP), blood products, Vitamin-K, TXA, rFVIIa, dextrose	GA, TXA, Vitamin-K, blood products	GA + TAP block, hemodynamic monitoring blood products, Vitamin-K, TXA, rFVIIa dextrose, mechanical ventilation, CRRT with endotoxin filter, ECMO	Neuraxial, hemodynamic monitoring, dextrose, TXA
Neonate	Female and male	Both female	Female	Female
Complications	Hemoperitoneum, coagulopathy/ DIC, HE, HTN emergency, hypoglycemia, AKI	Sepsis, coagulopathy	Bleeding/DIC, sepsis/MODS, AKI, ARDS, HE, hypoglycemia	Hypoglycemia, metabolic acidosis, bleeding

DM: Diabetes mellitus, HTN: Hypertension, GHTN: Gestational HTN, GDM: Gestational DM, POD: Postoperative day, LSCS: Lower segment cesarean section, GA: General anesthesia, TXA: Tranexamic acid, rFVIIa: Recombinant factor VII, DIC: Disseminated intravascular coagulation, CRRT: Continuous renal replacement therapy, ECMO: Extracorporeal membrane oxygenator. AKI: Acute kidney injury, MODS: Multiorgan dysfunction, ARDS: Acute respiratory distress syndrome, BMI: Body mass index, AFLP: Acute fatty liver of pregnancy, POG: Period of gestation, HE: Hepatic encephalopathy, ICU: Intensive care unit, CVC: Central venous catheter, IBP: Invasive blood pressure, TAP; Transversus abdominis plane block, IVF; In vitro fertilization

Table 2: "Swansea criteria" and its distribution among the cases

Number	Swansea criteria	Normal values	A1	A2	A3	A4
1	Clinical					
	Vomiting		−	−	+	+
	Abdominal pain		+	+	−	−
	Polyuria/polydipsia		−	−	+	−
	Encephalopathy		+	−	+	−
2	Laboratory					
	Elevated bilirubin	>14 µmol/L; >0.82 mg/dL	+	+	+	+
	Hypoglycemia	<4 mmol/L; <72 mg/dL	+	−	+	+
	Hyperuricemia	>340 µmol/L; >5.7 mg/dL	−	−	−	−
	Leukocytosis	>11,000/L	+	+	+	+
	Transaminitis	>42 IU/L	+	+	+	+
	Hyperammonia	>47 µmol/L; >66 µg/dL	−	−	−	−
	Increased creatinine	>150 µmol/L; >1.7 mg/dL	+	−	+	−
	Coagulopathy	PT>14 s/aPTT>34 s	+	+	+	+
3	Imaging					
	USG	Ascites/bright liver	+	+	+	−
	Liver biopsy	Microvesicular steatosis	−	−	−	−

USG: Ultrasonography, PT: Prothrombin time, aPTT: Activated partial thromboplastin time, +: present, -: absent

isolated transaminitis (5–10 times), coagulopathy (↑INR, ↓fibrinogen or commencing DIC), thrombocytopenia, and hypoglycemia defining the clinical features in up to 40%–50% cases.[3,4,8,12]

Majority burden of AFLP complications is owing to its severe multiorgan fatty infiltration, resulting in an acute kidney injury – 56%–85%, HE – 18%–50%, DIC – 28.3%, postpartum hemorrhage – 27.4%, multiple organ dysfunction syndrome (MODS) – 7%–28%, acute hepatic failure – 22%–47%, infections – 12.7%, and pulmonary edema/ARDS.[1,7,8,10] Pancreatitis and transient diabetes insipidus (elevated vasopressinase) are also commonly associated with this entity.[3,6] Maternal mortality occurs mainly due to hemorrhage, gastrointestinal bleeding, sepsis, aspiration, pancreatitis, or renal failure, whereas newborn morbidity/mortality may be due to hypoglycemia, metabolic acidosis, hepatic failure, and cardiomyopathy.[4,11]

Rarity of this entity renders diagnosis challenging, except for liver biopsy revealing microvascular infiltration of central and mid-zonal hepatocytes.[1,3,5] However, although the gold standard for diagnosis is liver biopsy, this procedure is not always necessary owing to its invasive nature in the setting of coagulopathy, therefore, Swansea diagnostic criteria (symptomatic and laboratory) are regarded as an alternative with positive and negative predictive value of 85% and 100%, respectively, as shown in Table 2.[1,3,5,7] Six or more criteria are required to diagnose AFLP when other possibilities more or less have been excluded from the study. Radiological investigations such as ultrasonography (USG), magnetic resonance and computed tomography imaging are supportive but not diagnostic. USG reveals hyperechoic fatty infiltration and ascites with low specificity/sensitivity and high false negatives.[4-5,7] However, in contrast a study revealed 46.3% patients of AFLP with classic bright liver/ascites on bedside USG (non-invasive point- of-care test).[7] Even in our case series, three out of four cases had ascites with bright liver on USG abdomen.

Liver diseases complicate 3% of all pregnancies with AFLP remaining a diagnosis challenge of exclusion, with several studies finding ways to differentiate other overlapping/coexisting conditions (even on liver biopsy).[3,5,12] HELLP and preeclampsia usually have high blood pressure with proteinuria without hypoglycemia, whereas preeclampsia is not jaundiced. Acute viral hepatitis in pregnancy also presents with fever, nausea, vomiting, fatigue, and jaundice, but the aminotransferase values are markedly elevated (>1000 U/L) and serology test will be positive, whereas IHCP can cause jaundice in any trimester and the itching is the characteristic symptom with raised serum bile acid levels.[2,4,12] Nausea and vomiting attributed to hyperemesis gravidarum may also produce hepatic dysfunction usually occurring in the first trimester, with drug-induced, autoimmune, and toxic causes to be ruled out.[11,12] Severe coagulopathy, jaundice, HE, ascites, hypoglycemia, and a mild-to-moderate elevation of transaminase levels with marked leukocytosis are the key features of AFLP.[2,5] Hence, pregnant women with abnormal liver functions require elaborate workup that includes blood/coagulation (viscoelastic testing), viral/autoimmune serology, drug/toxicology, and imaging to determine the etiology.[3,12]

Owing to the suggested pathophysiology and plethora of multiorgan complications in AFLP, early diagnosis, careful monitoring, comprehensive supportive intensive care, and immediate delivery with multidisciplinary approach remain the mainstay in the management, as it will break this chain of fatty acid circulation and diminish the hepatic stress[4,7] [Figure 1].

Interdisciplinary perioperative fetomaternal management by a team comprising anesthesiologists, gynecologists, hepatologists, intensivists, hematologists, transfusion physicians, nephrologists, and neonatologists should be assured.[6,12] Obstetric management requires prompt and safe delivery with individual consideration (fetal and maternal) of the method and timing of delivery. If spontaneous delivery is not imminent, induction for vaginal delivery without episiotomy (avoiding hemorrhagic complications) should be performed and cesarean section should be reserved in case of failure. In an emergency/coagulopathy setting, cesarean section may be harmful, however, in severe disease, it can help expedite delivery.[3,5,6,12]

There is no definitive literature for anesthetic management, however, it revolves around stabilization, monitoring, and extensive supportive care in the entire perioperative setting. Figure 1 gives an algorithm approach toward AFLP management. In the prepartum period, the fluid status, adequate blood products, and laboratory investigations need to be assessed, followed by correction of blood volume, hypoglycemia, coagulopathy, and extensive monitoring which may include invasive hemodynamic, viscoelastic, neurological monitoring for raised intracranial pressure, transoesophageal echocardiography, depth of anesthesia with neuromuscular monitoring to guide management and recovery.[3,5,6]

Intrapartum considerations include labor analgesia, choice of anesthesia for cesarean delivery, ongoing monitoring with fluid and hemodynamic management, control of coagulopathy with blood products and adjuncts such as TXA/Vitamin K/plasma, and avoidance of cerebral vasodilators or hepatotoxic drugs. Neuraxial anesthesia is typically favored due to low complications but involves risk with coagulopathy. GA may be the choice of anesthesia technique in presence of infection/sepsis and intracranial HTN/herniation, where neuraxial anesthesia is contraindicated. Conversely, GA can exacerbate hepatic dysfunction, intracranial HTN, and difficult airway. In addition, the transverse abdominal plane block can be adopted so as to decrease pain score with avoidance of opioids, sedation, or hepatotoxic acetaminophen/nonsteroidal anti-inflammatory drugs but may be used with caution for Local anesthetic systemic toxicity (LAST)/coagulopathy.[3]

Postpartum management includes ICU care with nutritional support, pain, and infection control (local/regional anesthesia

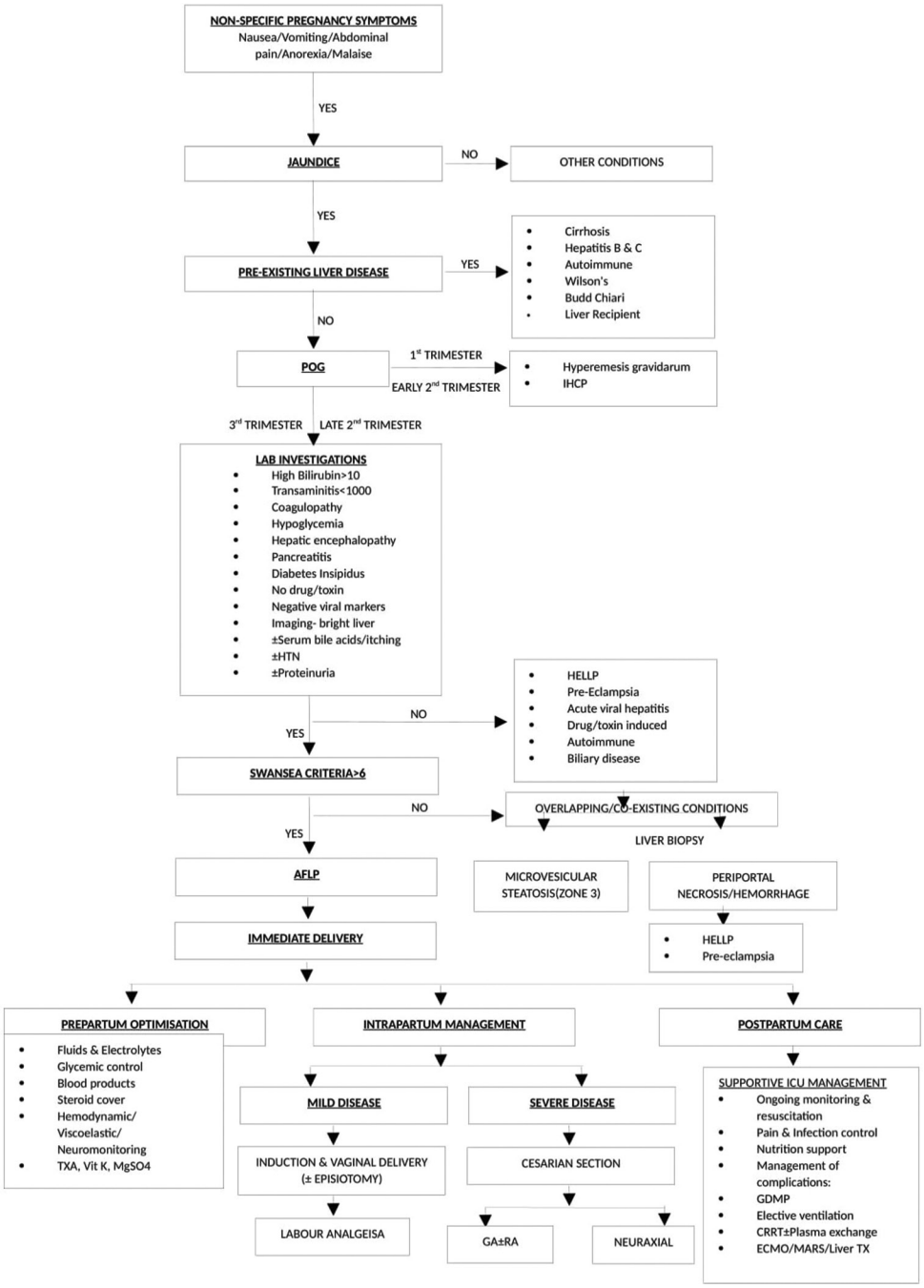

Figure 1: Algorithm approach toward acute fatty liver of pregnancy management. POG: Period of gestation, IHCP: Intrahepatic cholestasis of pregnancy, HELLP: Hemolysis, elevated liver enzyme, low platelets, AFLP: Acute fatty liver of pregnancy, TXA: Tranexamic acid, GDMP: Goal-directed medical protocol, CRRT: Continuous renal replacement therapy, ECMO: Extracorporeal membrane oxygenation, MARS: Molecular adsorbent recirculating system, TX: Transplantation, GA: General anesthesia, RA: Regional anesthesia

and low threshold for broad-spectrum antibiotics/antifungals as prophylaxis not recommended), continued hemodynamic/neurologic/laboratory evaluation and management of complications such as circulatory dysfunction, bleeding, hypoglycemia, sepsis, ARDS, and renal dysfunction. Additional ICU management such as plasma exchange with CRRT and ECMO has been shown to improve clinical recovery without mortality benefit. If hepatic dysfunction does not abate rapidly after pregnancy, molecular adsorbent recirculating system therapy, a nonbiologic albumin dialysis technique to stabilize liver function or liver transplantation should be a considered a lifesaving approach.[3,5]

Platelet/plasma/ATIII concentrate/rFVIIa transfusions and magnesium sulphate with its use in <32weeks prevents cerebral palsy and all cumulatively improve the clinical outcomes of mother and fetus.[3,5,7] Other considerations include fetal

tests (molecular long-chain 3 hydroxyacyl-CoA-dehydrogenase deficiency) and close monitoring for its manifestations, including hypoglycemia and fatty liver.[5]

Conclusion

AFLP is one of the rare liver diseases in pregnancy that remains a diagnostic conundrum with multitude of manifestations and overlapping conditions, an obstetric and perioperative emergency with high fetomaternal mortality. The absence of adequate data or directive guidelines makes the treatment ever-challenging, however, early diagnosis, prompt delivery, and aggressive perioperative multidisciplinary supportive care remain the cornerstone of management.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Meng Z, Fang W, Meng M, Zhang J, Wang Q, Qie G, *et al.* Risk factors for maternal and fetal mortality in acute fatty liver of pregnancy and new predictive models. Front Med (Lausanne) 2021;8:719906.
2. Ziki E, Bopoto S, Madziyire MG, Madziwa D. Acute fatty liver of pregnancy: A case report. BMC Pregnancy Childbirth 2019;19:259.
3. Naoum EE, Leffert LR, Chitilian HV, Gray KJ, Bateman BT. Acute fatty liver of pregnancy: Pathophysiology, anesthetic implications, and obstetrical management. Anesthesiology 2019;130:446-61.
4. Yadav A, Sharma C, Kathpalia SK, Singh SS. Acute fatty liver of pregnancy: A clinical dilemma. Indian J Community Fam Med 2020;6:171-4.
5. Hadi Y, Kupec J. Fatty liver in pregnancy. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2023.
6. Yemde A Jr., Kawathalkar A, Bhalerao A. Acute fatty liver of pregnancy: A diagnostic challenge. Cureus 2023;15:e36708.
7. Chang L, Wang M, Liu H, Meng Q, Yu H, Wu YM, *et al.* Pregnancy outcomes of patients with acute fatty liver of pregnancy: A case control study. BMC Pregnancy Childbirth 2020;20:282.
8. Fesenmeier MF, Coppage KH, Lambers DS, Barton JR, Sibai BM. Acute fatty liver of pregnancy in 3 tertiary care centers. Am J Obstet Gynecol 2005;192:1416-9.
9. Ademiluyi A, Amakye DO, Jackson N, Betty S. Acute fatty liver of pregnancy. Am J Case Rep 2021;22:e933252.
10. Rolfes DB, Ishak KG. Liver disease in pregnancy. Histopathology 1986;10:555-70.
11. Dey M, Reema K. Acute fatty liver of pregnancy. N Am J Med Sci 2012;4:611-2.
12. Maier JT, Schalinski E, Häberlein C, Gottschalk U, Hellmeyer L. Acute fatty liver of pregnancy and its differentiation from other liver diseases in pregnancy. Geburtshilfe Frauenheilkd 2015;75:844-7.