Elective Caesarean Delivery in a Primigravida with CPT II Deficiency: A Multidisciplinary Anaesthetic Approach

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Carnitine Palmitoyl Transferase II Deficiency (CPT II) is a rare disorder of long-chain fatty acid oxidation

- Three forms: adult, infantile, perinatal (autosomal recessive) Adult form: relatively benign; symptoms triggered by stress (e.g. intense exercise)
- Limited evidence on anaesthetic management
- Reported risks: malignant hyperthermia, propofol infusion syndrome, prolonged neuraxial blockade
- Metabolic stress may cause: hypoglycaemia, rhabdomyolysis, hyperkalaemia, acidosis, myoglobinuria, acute kidney injury

Background

- 33-year-old primigravida with Carnitine Palmitoyl Transferase II Deficiency
- Diagnosed at 21 after severe myalgia + rhabdomyolysis → ICU admission (dialysis required)
- Diagnosis confirmed by skin biopsy
- Follow-up via National Centre for Inherited Metabolic Disorders Stable for 12 years; one disease-related admission 2 years prior

Investigations & Planning

Baseline: Vitals normal; weight 99 kg; airway unremarkable

No prior GA, no FHx anaesthetic complications, no allergies

Reported prolonged effect of local anaesthetic

Medications: Calcichew, MCT oil 15 mL BD, L-carnitine 1 g BD, pregnancy supplement

Low-fat, high-carbohydrate diet

Investigations: Echo + liver ultrasound normal

Anaesthetic plan (MDT):

Regional preferred (↓ metabolic stress)

If GA: avoid propofol infusions, lipid emulsions, suxamethonium; use non-triggering agents

Minimise fasting; IV 20% dextrose @ 90 mL/hr

Metabolic plan:

Pregnancy/labour: fasting <3 h, high-carb drinks (SOS25), restrict LCTs, continue MCT if stressed

Pre-op (2 days): LCT ≤40 g/day; ↑ intake to ~2690 kcal/day

Monitoring:

Peri-op: glucose, electrolytes, CK, LFTs, FBC, coagulation

Postpartum: acylcarnitine profile (day 3)

Postpartum:

Nutrition guided by CK; return to baseline diet by day 3

Interventions & Intra-operative Course

- IV 20% dextrose commenced; first on list (↓ fasting)
- MH precautions in place
- **Anaesthesia:** combined spinal-epidural (CSE)

Intrathecal: prilocaine 2%, fentanyl, morphine

Epidural: lidocaine 2% supplementation

- Arterial line inserted pre-induction
- Caesarean delivery uneventful - EBL 920 mL; haemodynamically stable
- Motor block resolved at ~10 hours
- Early oral intake in PACU + continued dextrose

Postoperative Course & Outcome

- HDU admission postoperatively
- Prolonged motor block (~10 hrs) → full recovery, no deficits
- CK >3000 U/L (associated with diarrhoea) → resolved with ↑ caloric intake
- Glucose stable throughout
- L-carnitine increased to 3 g daily
- IV potassium supplementation continued
- Discharged: HDU day 2; home day 7, well

Discussion & Conclusion

- Highlights importance of MDT perioperative planning in Carnitine Palmitoyl Transferase II Deficiency
- CSE with prilocaine provided effective, metabolically safe anaesthesia
- Prolonged motor block anticipated; no adverse sequelae
- Post-op metabolic stress (diarrhoea) → CK rise → required nutritional optimisation
- GA contingency: MH precautions; avoid propofol, lipid emulsions, suxamethonium
- Coordinated approach → favourable maternal and neonatal outcome

References:

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