



Introduction

Glutaric Aciduria Type 1 (GA1)

- Rare autosomal recessive metabolic disorder
- Caused by: Deficiency of glutaryl-CoA dehydrogenase → accumulation of glutaric and 3-hydroxyglutaric acids.¹
- Impaired glucose metabolism in the brain
- Catabolic stress → encephalopathic crisis
- Acute decompensation → irreversible brain injury and dystonic cerebral palsy.

Management of GA1

- Protein restriction, L-carnitine supplementation (enhances glutaryl-CoA excretion)
- Avoid catabolism during metabolic stress

Pregnancy

- Increased metabolic demand (catabolic stress of labour and fasting) → metabolic decompensation.

Reports of successful induction of labour (IOL) or emergency caesarean delivery are limited.

Case

Preparation

POAC

- 30-year-old primigravida POAC at 38 weeks’ gestation with GA1
- Examination unremarkable
- No prior exposure to GA
- Medications: L-Carnitine 1g QDS, GA Express®15 QDS and a pregnancy-specific supplement.
- Adhered to a low-protein diet.

Plan by MDT (obstetrics, anaesthesiology, metabolic medicine, and dietetics)

- Induction of labour (IOL) at 39+1 weeks’ gestation
- Regional anaesthesia: Limit catabolic stress and reduce risk of metabolic decompensation
- Certain agents to avoid: Hartmann’s, prolonged propofol infusions (potentially harmful to mitochondrial metabolism).
- Peri-operative fasting minimised: Reduce the risk of catabolic crisis.
- Individualised metabolic plan for both oral and parenteral nutritional supplementation for IOL:
 - Increase calorie consumption to 120% of usual energy requirements
 - Parenteral nutrition: Intravenous Dextrose 20% with 7mLs L-Carnitine (1g/5mLs) at 74mLs/hr for 24hours and Intravenous Intralipid 20% at 25mLs/hr for 24 hours
- PICC insertion prior to admission for IOL
- Serial blood gas analyses: monitor for lactatemia.

Management

Admission for IOL

- Admitted to delivery suite: Oxytocin infusion commenced.
- 0.9% sodium chloride at 125mLs/hr to prevent dehydration.
- Early epidural analgesia: Minimise stress-induced catabolism.
- Baseline lactate on arrival 1.2 mmol/L → 1.6 mmol/L (10 hours later)
- Parenteral nutrition commenced due to rising lactate and poor oral intake
- Oxytocin infusion titrated up to 32 mL/hr.
- Oedematous and positive fluid balance of 2 L → 0.9% sodium chloride IVI stopped.

Emergency Caesarean Section (CS)

- Indication: FTP
- Epidural block T10 b/l → top-up 240 mg lidocaine, 60 mcg adrenaline and 100 mcg fentanyl → Block T4 b/l
- Parenteral nutrition continued throughout CS. No additional IVF administered.
- Uncomplicated CS
- EBL 600mLs

Post op

- HDU
- Lactate peak 2.9 mmol/L → normalised in 12 hours.
- Parenteral nutrition continued
- Gradual reintroduction of a low-protein diet under dietetic supervision and tapering of parenteral
- Neurological examination was normal and she suffered no metabolic complications.

Key Points

1. Highlights complexity of managing GA1 in pregnancy
2. Multidisciplinary involvement is essential in managing metabolic disorders in pregnancy to optimise patient outcomes.
3. Regional anaesthesia is preferred to minimise catabolic stress and reduce the risk of metabolic decompensation.
4. Interpretation of lactate in labour in a patient with GA1 requires caution as modest rises are common in labour and peri-operatively. (In healthy parturients, lactate <4 mmol/L is usually physiological.²)
5. Ensure frequent fluid and nutrition assessments and adjustments based on clinical status.

1. Li Q, Yang C, Feng L, Zhao Y, Su Y, Liu H, et al. Glutaric Acidemia, Pathogenesis and Nutritional Therapy. Front Nutr. 2021;8:704984.

2. Dockree S, O’Sullivan J, Shine B, James T, Vatish M. How should we interpret lactate in labour? A reference study. Bjog. 2022;129(13):2150-6.