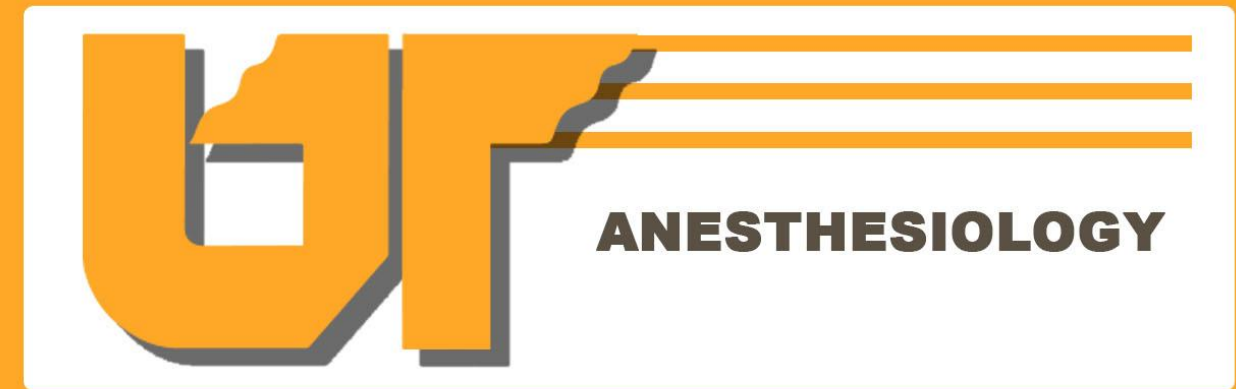




Anesthetic Management of a Parturient with Carnitine Palmitoyltransferase II Deficiency

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Background:

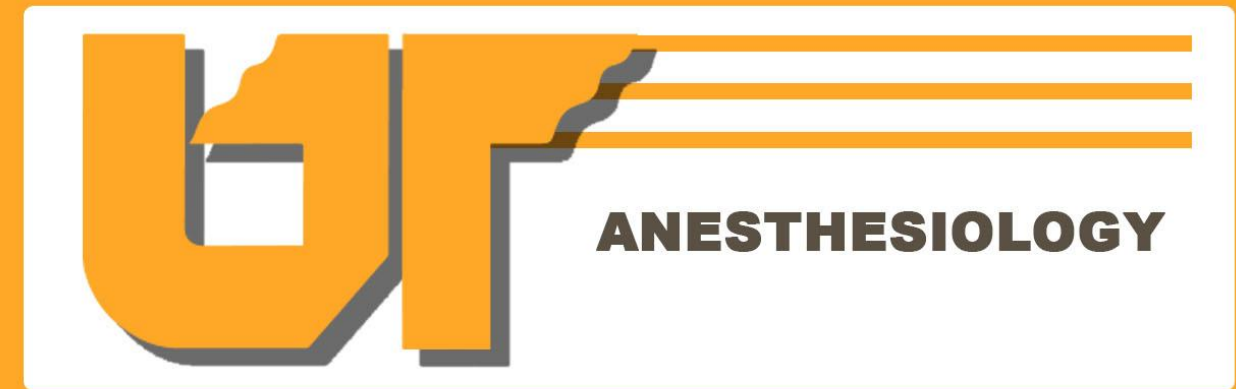
- Carnitine palmitoyltransferase II (CPT II) deficiency is an autosomal recessive disorder of long-chain fatty-acid oxidation within the mitochondria
- 3 known forms
 - 1. Neonatal form (often lethal)
 - 2. Severe infantile form
 - 3. Adult myopathic form (most common)
- Patients with myopathic form will experience muscle pain due to rhabdomyolysis and dark-colored urine due to myoglobinuria when exposed to known triggers
 - Stress
 - Physical exertion
 - Fasting
 - Infections
- If rhabdomyolysis is severe it can result in acute renal failure and even death.¹



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Case:

- 34-year-old G3P1 at 37.2 presents for induction of labor. History significant for CPT II deficiency.
 - She was hospitalized the year prior with significant muscle pain after exercise with severely elevated CK levels.
 - Subsequent neurology consultation with metabolic myopathy panel revealing CPT II deficiency.
- On L&D the patient was initiated on D5LR @ 125cc/hr.
- An epidural was placed and a plan discussed for etomidate and rocuronium RSI if emergency cesarean delivery was indicated given her hx.
- The patient had an uncomplicated spontaneous vaginal delivery and was discharged home the following day with a healthy neonate.

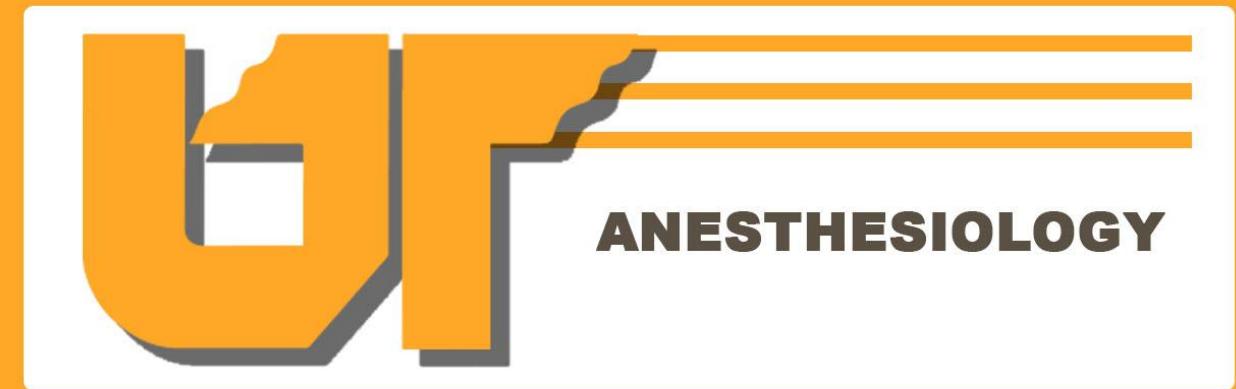




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Discussion/Learning Points:

•Glucose containing infusions

- Glucose containing infusions are imperative in the perinatal period with frequent glucose monitoring. Patients with impaired ability to utilize long-chain fatty acids for energy rely on carbohydrate sources. When stressors are present (fasting, labor, surgical stress), the body will resort to muscle breakdown when insufficient carbohydrates are present.

•Mitigate stress response

- Early epidural is recommended to prevent the stress response associated with labor.

•Stat section planning

- Our institution's standard RSI with propofol and succinylcholine would be inappropriate for this patient. Propofol is dissolved in 10% long-chain fatty acids and thus should be avoided in patients with compromised fatty acid β -oxidation. It is generally recommended that succinylcholine be avoided given the risk of exacerbating rhabdomyolysis. RSI with etomidate and rocuronium would be most appropriate with maintenance of general anesthesia using volatile/nitrous oxide.

References:

- 1.Wieser T. Carnitine Palmitoyltransferase II Deficiency. 2004 Aug 27 [updated 2019 Jan 3]. In: Adam MP, Feldman J, Mirzaa GM, Pagon RA, Wallace SE, Amemiya A, editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993–2025. PMID: 20301431.
- 2.Yu HK, Ok SH, Kim S, Sohn JT. Anesthetic management of patients with carnitine deficiency or a defect of the fatty acid β -oxidation pathway: A narrative review. Medicine (Baltimore). 2022 Feb 18;101(7):e28853. doi: 10.1097/MD.00000000000028853. PMID: 35363184; PMCID: PMC9282055.

