

Postpartum Myonecrosis in a Patient with VLCAD - The Epidural Blame Game Continues...

Marshal Lu, M.D., Erik Romanelli, M.D., Yelena Spitzer, M.D.

Very-long-chain acyl-CoA dehydrogenase deficiency (VLCAD)

- Genetic disorder causing impaired mitochondrial beta-oxidation of long and very-long chain fatty acids
- Three phenotypes
 - Severe neonatal form
 - Childhood hypoketotic hypoglycemic form
 - Adult-onset episodic myopathic form
- Most common - Adult-onset episodic myopathic form
 - Muscle cramps, exercise intolerance, and intermittent rhabdomyolysis

36F presented for induction of labor of her third pregnancy

- Known ACADVL gene mutation for mitochondrial VLC acyl-coenzyme A dehydrogenase
 - Exercise intolerance and frequent muscle cramping at home
- Pregnancy 7 years ago was complicated by rhabdomyolysis (CK 5,596 U/L) and inflammation and severe pain to the left thigh and buttocks
 - CT/MRI suggestive of a compressive injury with possible myonecrosis of left gluteus and proximal vastus lateralis



Current pregnancy

- Precautions taken:

- Frequent glucose monitoring
- Baseline CK was drawn: 187 U/L
- Oral and IV glucose-containing solutions and fluids
- Frequent positional changes
- CSE performed

- Labored for 48h → cesarean section for arrest of labor

- Postpartum, she had left hip and thigh pain NOT associated with increased CK

- Symptoms improved and she was discharged home with her child

High risk of rhabdomyolysis during states of increased metabolic demand (LABOR)

- Other associated risk factors:
hypoglycemia, pain, stress
- Major complication = kidney damage

Suggested precautions:

- Expeditious labor to reduce energy demand
- Avoid hypoglycemia - oral and IV dextrose
- Hydration for renal protection
- Normothermia: avoid shivering / increased energy use in hyperthermia
- Epidural analgesia to reduce pain and stress
- Frequent turning to avoid compressive muscular injury

What if you need to provide general anesthesia?

Avoid the following:

- Lipids: propofol, formulations of etomidate in lipid emulsion
- Succinylcholine: can cause myalgia
- Volatile anesthetics? enflurane has been shown to increase free fatty acids, but volatile anesthetics have been used without problems

May use these drugs:

Ketamine, opioids, benzodiazepines, barbiturates, dexmedetomidine, and non-depolarizing neuromuscular blocking drugs

We suggest RSI with ketamine and rocuronium with TIVA maintenance with ketamine, dexmedetomidine, and remifentanyl