Volatile anesthesia for a pediatric patient with very-long-chain acyl-coenzyme A dehydrogenase deficiency

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Introduction: Very long-chain acyl coenzyme A dehydrogenase deficiency (VLCAD) is a rare, autosomal recessive disorder of fatty acid metabolism that places the patient at risk of hypoglycemia, cardiomyopathy and liver failure. Volatile anesthetics have historically been avoided because of case reports associating them with catabolic crises, but a recent literature review suggests the reports of rhabdomyolysis in VLCAD patients receiving general anesthesia were either in the context of prolonged fasting or not associated with volatile anesthetic agents.

Case: A 3-year-old 18.5-kg boy with a history of infantile-type VLCAD and G-Tube dependence presented to our institution for tonsillectomy.
  • Prior anesthetic for G-tube included ketamine and remifentanil though difficult to maintain adequate anesthetic depth
  • Consulted Pediatric Genetics service and planned volatile-based anesthetic with minimal fasting time and emotional distress
  • G-tube feeds were held 8 hours prior to surgery and clear liquids were continued until 2.5 hours preoperatively
  • Upon arrival an IV was placed and a 10% dextrose infusion was started at a maintenance rate

Perioperative Course:
  • Premedication with IV midazolam (0.04 mg/kg)
  • Inhalation induction with sevoflurane/N2O/Air
  • #2 flexible LMA placed, spontaneous ventilation
  • Maintenance with 3% sevoflurane, 50% O2/Air
  • 10 mL/kg 0.9% NS along with D10/NS at maintenance rate
  • Dexamethasone and ondansetron (both 0.1 mg/kg)
  • Hydromorphone (0.01 mg/kg) and dexametomidine (0.5 mcg/kg)
  • LMA removed under deep anesthesia, calm during PACU stay
  • Tolerating G-tube feeds, discharged POD #1

Discussion: VLCAD results from mutation of the ACADVL gene and is subdivided into neonatal (severe), infantile, and late-onset (episodic) forms depending on severity and the degree of enzyme deficiency. Caloric insufficiency and emotional distress may precipitate severe catabolism and rhabdomyolysis. Lipid exposure leads to accumulation in tissues and causes long-term organ dysfunction.
  • Contraindicated medications: Propofol (10% soybean oil emulsion) and succinylcholine (rhabdomyolysis risk)
  • Safe medications: Opiates, dexametomidine,ketamine, NDMRs

There are published case reports of volatile anesthetics safely used in adults with VLCAD. Prior case reports of volatile-associated rhabdomyolysis in pediatric VLCAD patients were likely due to inadequate caloric supplementation rather than the anesthetic agent.

Learning Points:
1. VLCAD is a rare disorder of fatty acid metabolism that places the patient at risk of hypoglycemia, cardiomyopathy and liver failure. Caloric insufficiency and emotional distress may precipitate severe catabolism and rhabdomyolysis.
2. Avoidance of fasting through careful preoperative planning and IV glucose supplementation is critical for the successful perioperative management of patients with VLCAD.
3. Volatile anesthetics are likely safe in VLCAD and may have a protective effect against catabolism and rhabdomyolysis.

References: