Anesthetic Management for a Pediatric Patient with Very Long-Chain Acyl-Coenzyme A Dehydrogenase Deficiency

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BACKGROUND

Very Long-Chain Acyl-Coenzyme A Dehydrogenase Deficiency (VLCADD) is an autosomal recessive disorder involving the initial steps in fatty acid metabolism with potential for hypoglycemia, rhabdomyolysis, liver failure, fatty acid intermediate toxicity, and mitochondrial dysfunction. There is a paucity of articles related to the anesthetic management of VLCADD. Authors suggest avoidance of propofol and volatile anesthetic.[1,2,3] Opioids, NSAIDS, and regional anesthesia have been administered safely. We present a contemporary alternative approach to the anesthetic management of a pediatric patient with VLCADD with the administration of etomidate, remifentanil, ketamine, rocuronium, sugammadex, dexmedetomidine and fentanyl.

CASE

A 14 year old patient (80 kg) with childhood VLCADD presented for port insertion. PMHx included multiple hospitalizations for recurrent rhabdomyolysis due to VLCADD exacerbations with food poisoning, menses, exercises and dehydration. Plasma creatinine kinase (CK) levels ranged 30,000 - 57,000 U/L during episodes. PSHx was significant for G-tube placement. Medications included: paroxetine hydrochloride and levocarnitine.

Anesthesia Management

The patient was admitted the day prior to surgery and anesthesia was consulted. Blood was sent for baseline levels of liver/kidney function, electrolytes, glucose, and CK. All results were WNL except for a CK (520 U/L). IVF with D10% ½NS at 125 ml/h was initiated. The patient refused local anesthesia/sedation. General anesthesia was planned and discussed with the family.

On the day of surgery, IVF with D10% LR 200 ml/h (glucose 4 mg/kg/min) was administered. The patient was premedicated with midazolam 2 mg IV. General anesthesia was induced with morphine 10 mg IV slowly, lidocaine 50 mg IV, etomidate 24 mg IV, and rocuronium 80 mg IV. TIVA was maintained with a remifentanil infusion at 0.1-0.2 mcg/kg/min and ketamine 50 mg lasting 102 minutes. Sugammadex 160 mg IV antagonized the NM block. Fentanyl

CASE CONT'D

(40 mcg IV) and dexmedetomidine (20 mcg IV) were administered during tracheal extubation. The PACU course was unremarkable. Intra-operative and post-operative laboratory results showed no signs of hypoglycemia, hyperkalemia or CK elevation (Table 1). The patient was discharged home the following day without any complications.

TABLE 1: PRE-DURING-POST ANESTHESIA LABORATORY RESULTS IN PEDIATRIC PATIENT WITH VLCADD

Location	K^+(mEq/L) Normal (3.5-5.1)	Glucose (mmol/L) Normal (3.6-6.1)	CK (Units/L) Normal (47-312)
Pre-Operative	3.6	5.3	520
Intra-Operative	3.1	11.1	-
Post-Operative	3.9	11.4	359

DISCUSSION

VLCADD is associated with abnormal fatty acid metabolism especially during stress. The literature has a paucity of articles on the anesthetic management for patients with VLCADD. Glucose infusion to avoid hypoglycemia and avoiding dehydration are important. Opioids, benzodiazepine, NSAIDS, and local anesthetic have been safely used. In view of the lack of reports in the literature for the anesthetic management for pediatric patients with VLCADD, we present a successful alternative anesthetic management (etomidate, remifentanil, ketamine, rocuronium, sugammadex, dexmedetomidine and fentanyl) never previously reported.

REFERENCES

1. Steiner LA, Studer W, Baumgartner ER, Frei FJ. Perioperative management of a child with very-long-chain acyl-coenzyme A dehydrogenase deficiency. Paediatr Anaesth. 2002 Feb;12(2):187-91.
2. Kleemann PP, Jantzen JP, Fenner R, et al. Preoperative increase in the plasma concentration of free fatty acids during minor elective interventions using a conventional anesthesia technic with enflurane. Anaesthesist. 1986 Oct;35(10):604-8.
3. Welsink-Karssies MM, Polderman JAW, Nieveen van Dijkum EJ, Hermanides J, et al. Very Long-Chain Acyl-Coenzyme A Dehydrogenase Deficiency and Perioperative Management in Adult Patients. JIMD Reports. 2017;34:49-54.