Introduction:

- Mitochondrial beta-oxidation of fatty acids represents a major source of energy under fasting and stressful conditions to the body.
- Errors in this process, especially long chain fatty acids with >14 carbons, lead to accumulation of harmful acyl-CoA and acylcarnitine that can promote dysrhythmias (1,2).
- The first case of very-long-chain acyl-CoA dehydrogenase deficiency (VLCADD) was documented in 1993 (3).
- VLCADD is an autosomal recessive disease with an incidence of 1:31,500 births. It can be diagnosed in newborns with elevation of c14:1 acylcarnitine level, fibroblasts VLCAD activity analysis, or genetic analysis (4,5).
- This disease presents a challenge for perioperative management of patients since fasting or perioperative stress can lead to an increase in energy requirement and utilization of fatty acid oxidation, resulting in episodes of metabolic acidosis, hypoketotic hypoglycemia, myocardial or skeletal dysfunction (4).
- Additionally, the commonly used IV anesthetic propofol and inhalational agent sevoflurane were previously reported to cause metabolic acidosis in these patients (6,7). We recently encountered a 2-month old patient with VLCADD for unilateral cleft lip repair.

Case Report:

- TC was a two month-old full term 4.5 kg baby boy with past medical history significant for VLCADD, previously diagnosed via genetic testing.
- On the day prior to surgery, he was admitted to the pediatric floor for IV fluid hydration with glucose.
- In the preoperative area, TC received 0.5 mg IV of midazolam prior to being brought back to the operating room.
- For anesthesia induction, 0.1 mg/kg vecuronium, 2 mcg/kg fentanyl and 0.1 mg/kg midazolam IV were titrated to effect prior to successful intubation with an oral RAE endotracheal tube.
- Anesthesia was maintained with remifentanil and dexmedetomidine infusions with intermittent 0.05 mg/kg boluses of midazolam. BIS monitor was also used to assure adequate anesthetic level. 200 mg L carnitine IV was given one hour into the case per recommendation from Medical Genetics Service. The intraoperative course was uneventful.
- Pt was reversed with glycopyrrolate, neostigmine, and successfully extubated wide awake shortly after surgery ended. Dexmedetomidine and D10 ¼ NS infusions were continued into the PACU.
- Serum glucose was 105 in PACU and all subsequent values were normal. Creatinine Kinase levels were normal POD #1, effectively ruling out any postoperative rhabdomyolysis. Perioperative course on the floor was uneventful. Pt was discharged to home on POD #1.

Discussion:

- In this patient with VLCADD, the goal of our anesthetic management was to avoid any increase in physical stress associated with fasting, emotional stress in the preoperative area, and intraoperative surgical stress.
- To minimize decompensation from fasting, the patient was admitted to the hospital the day before for IV fluid hydration with glucose. In the preoperative area, patient was given midazolam IV to help with emotional stress.
- There were reports of usage of propofol and sevoflurane in patients with VLCADD with mixed results (6). Propofol formulary contains long chain fatty acids and is known to cause entry impairment of long chain fatty acids into the mitochondria for oxidation, leading to similar presentations seen in VLCADD patients: arrhythmia, metabolic acidosis, cardiac failure, fatty liver, rhabdomyolysis, and renal failure (8). On the other hand, usage of inhalational agents was associated with elevation of fatty acids as reported by Kleeman et al. (7).
- To minimize the risk of fatty acids accumulation, we decided to use remifentanil and dexmedetomidine infusions with intermittent midazolam boluses for maintenance since remifentanil and midazolam were used previously with no complications (9,10).
- Given the risks of myopathy associated with errors in mitochondrial beta oxidation of fatty acids, there might be a possibility of prolonged paralysis with muscle relaxant usage. However, as previously reported by Schmidt et al., usage of midazolam and vecuronium were not associated with prolonged paralysis in VLCADD and mitochondrial disease patients (10).
- We were able to reverse and successfully extubated the patient at end of case. Dexmedetomidine infusion was continued into the PACU to decrease stress associated with anesthesia emergence.

Figure 1: Fatty acid metabolism pathway

Figure 2: Intraoperative anesthesia record

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