

[NM-343] Perioperative anesthetic management of a patient with very-long-chain acyl-coa dehydrogenase deficiency for unilateral cleft lip repair

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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). Very-long-chain acyl-coa dehydrogenase deficiency (VLCADD) presents a challenge for perioperative management of patients since the commonly used IV anesthetics such as propofol and inhalational anesthetics were previously reported to cause metabolic acidosis and rhabdomyolysis (3,4). Here, we report the anesthetic management for a 2-month old patient with VLCADD that was diagnosed via genetic testing for unilateral cleft lip repair. In the preoperative area, the child was sedated with IV midazolam. Fentanyl and midazolam IV were titrated to effect in the OR for induction. Muscle relaxation was achieved with vecuronium prior to oral intubation. Remifentanyl and dexmedetomidine infusion were started afterward for maintenance with BIS monitor to assure adequate anesthetic depth. A dextrose infusion was also started preoperatively and continued throughout the case to prevent episodes of hypoglycemia. Intraoperative course was uneventful. Patient was reversed with glycopyrolate, neostigmine, and successfully extubated wide awake shortly after surgery ended. The main goal in caring for patients with VLCADD is to avoid any physical and emotional stress that can lead to accumulation of fatty acids that are harmful to the body as well as possible triggering anesthetic agents that may increase the risk of metabolic acidosis or rhabdomyolysis (3,4). To minimize accumulation of exogenous fatty acids, we decided to go with total IV anesthetic for maintenance. After careful consideration, we chose remifentanyl and dexmedetomidine infusion with intermittent midazolam boluses since remifentanyl and midazolam were used previously with no complications (5). Using the technique presented above, we were able to keep the patient safe throughout the perioperative course for his cleft lip surgery. Additionally, our case demonstrated the safe usage of dexmedetomidine infusion in patients with VLCADD that had not been reported before.

#### References:

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