Anesthetic Challenge in a case of Propionic Acidemia

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BACKGROUND

Propionic Acidemia is a rare disease of the propionyl-CoA carboxylase that occurs in 1 every 100,000 cases. This population is unable to synthesize succinyl-CoA from propionyl-CoA, and present clinically with metabolic acidosis, hyperammonemia, hypotonia, and failure to thrive. Thus, anesthetic management of patients with propionic academia is challenging because several common anesthetic agents can trigger severe metabolic acidosis. To date, only 10 individual anesthetic cases are reported involving patients with propionic acidemia. We present with a case of a 5 week old female with propionic acidemia scheduled for laparoscopic gastrostomy and gastric tube placement.

Case Description: Preoperative Management

Patient was a full term infant born without family history of genetic diseases. Initial diagnosis was made at 5 days of life when she presented with severe metabolic acidosis and hyperammonemia. Urine organic acid profile was positive for 3-hydroxypropionic acid, methylcitric acid, and 3-hydroxyvaleric acid, but separate methylmalonic acid assay was within normal range. Specific mutation was identified on chromosome 3q22.1q25.2 which contains the PCCB gene, Patient was subsequently diagnosed as propionic acidemia.

Case Description: Intraoperative Management

On the day before surgery, the patient had been stabilized on room air without dialysis. She had tolerated nasogastric tube feeds with Similac Advance, Propimex-1, and Pro-Phree. However, she demonstrated significantly decreased oral motor skills as her feeding is exclusively through nasogastric tube, and was scheduled for gastrostomy tube placement. Her physical exam was unremarkable, and her intravenous fluid consisted of D10W at 20mL/hr.

Preoperative anesthetic plan include the following:

- Avoiding propofol, which was high in polyunsaturated fats.
- Avoiding lactate ringer, since she is prone to lactic acidosis.
- Avoiding succinylcholine, atracurium, mivacurium, since odd-chain organic molecules that are metabolized by ester hydrolysis are ultimately metabolized to propionyl-CoA.
- Avoiding ibuprofen, naproxen and other NSAIDS derived from propionic acid.
- Delay extubation until regained full muscle strength.

In the operating room, standard ASA monitors were used. Planned rapid sequence induction was performed with ketamine 0.5mg/kg and rocuronium 0.1mg/kg. Patient rapidly desaturated but recovered with manual ventilation. She was tracheally intubated successfully on the second attempt with a 3.0 cuffed tube as the 3.5 ETT was too large to pass her vocal cords on the first attempt. Cuff leak was noted at 20cmH2O, and the endotracheal tube was taped at 9.5cm at the lips. N2O, O2, and desflurane were used for anesthetic maintenance. Her maintenance fluid was D10W at 20mL/hr and NS available for resuscitation. She received acetaminophen intraoperatively for analgesia. Ultimately, patient received reversal of muscle relaxants, and was extubated with good muscle tone and spontaneous breathing. She recovered uneventfully in the PACU.

Case Description: Intraoperative Management

Figure 1: Metabolic Pathway leading up to Propionic Acidemia

CONCLUSIONS

Patient with propionic acidemia presents challenges to anesthesiologists that required detailed avoidance of certain anesthetic medication to avoid triggering episodes of metabolic acidosis. This case report adds to a small but growing case reports of successful anesthetic management to this unique population.