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Case Report

Tonsillectomy in a pediatric patient with propionic acidemia: Anesthesia management and potential perioperative challenges

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ABSTRACT

Propionic acidemia (PA) is a rare metabolic disorder caused by a deficiency in the propionyl-CoA carboxylase enzyme, leading to the accumulation of toxic metabolites. Surgical procedures pose a risk of metabolic decompensation in patients with PA, requiring careful anesthesia management. A four-year-old male patient with propionic acidemia was scheduled for tonsillectomy due to recurrent upper respiratory tract infections. Midazolam was administered for premedication, and anesthesia induction was achieved with thiopental and rocuronium. Anesthesia maintenance was provided with sevoflurane, and fentanyl was used for analgesia. Postoperative pain management included paracetamol. The procedure was completed without complications, and the patient was transferred to the ward in stable condition. Anesthesia management in PA patients should focus on maintaining metabolic stability. This case demonstrates that with proper preoperative preparation, careful anesthesia management, and close perioperative monitoring, surgical interventions can be safely performed in pediatric patients with PA.

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1. Introduction

Propionic acidemia (PA) is a rare autosomal recessive metabolic disease caused by deficiency or dysfunction of the enzyme propionyl-CoA carboxylase [1]. It occurs as a result of pathogenic mutations in the PCCA or PCCB genes. These genes encode the enzyme propionyl-CoA carboxylase, which is found in mitochondria. Propionyl-CoA carboxylase is involved in the catabolism of branched-chain amino acids, the process by which proteins are broken down for cellular metabolism. Enzyme deficiency results in the accumulation of propionic acid and related metabolites (propionyl-CoA, 2-methylcitrate, 3-OH-propionate). Accumulation of these toxic metabolites may lead to secondary mitochondrial dysfunction, resulting in progressive organ damage [2]. This prevents the conversion of propionyl-CoA to methylmalonyl-CoA and leads to the accumulation of toxic metabolites, resulting in clinical symptoms such as metabolic acidosis, ketosis and hyperammonemia [1]. Surgical procedures in patients with PA require careful management of anesthesia, as they can cause serious complications due to increased metabolic stress [3].

Significant metabolic abnormalities, including low levels of free carnitine, elevated C3 propionyl carnitine, and varied amino acid imbalances may be present [4]. Main challenges related to this rare case are higher incidence of cardiac manifestations like dilated cardiomyopathy due to accumulation of toxic metabolites and in-

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flammation in the heart muscle, long QT syndrome, life threatening arrythmias [5] white matter involvement and ventricular dilatation, acute decompensation caused by the catabolic stress of surgery and liver disease as propionyl-CoA carboxylase is primarily expressed in the liver [6].

Tonsillectomy is a common surgical procedure performed in children, but anesthetic management in patients with metabolic diseases such as PA poses challenges. This case report discusses the anesthetic management during tonsillectomy of a 4-year-old boy with PA and the challenges encountered in the perioperative period.

2. Case Presentation

A four-year-three-month-old, 13-kg male child diagnosed with propionic acidemia was scheduled for tonsillectomy due to frequently recurring upper respiratory tract infections and was taken into surgery. The patient's PA diagnosis was made on the third day of life and he was regularly followed up by a pediatric metabolic diseases specialist. The patient is on a special protein-restricted diet and is taking carnitine, carglutamic acid and vitamin B12 supplements. The patient, who had no history of seizures and was metabolically stable, was prepared for surgery.

During the premedication phase, 1.3 mg midazolam was administered intravenously to the patient and he was taken to the operation room. The patient's vital signs were monitored and his pulse was measured as 123/min, SpO2 was 94% and blood pressure was 98/54 mmHg. Fingertip blood sugar was measured as 137 mg/dL. 65 mg thiopental was administered for anesthesia induction and neuromuscular blockade was achieved with 9 mg rocuronium. For analgesia, 15 mcg fentanyl was used and the patient was intubated with a size 4.5 cuffed endotracheal tube.

Anesthesia was maintained with sevoflurane MAC (minimum alveolar concentration) 1.3 during the operation and the surgery lasted 35 minutes. Estimated blood loss was 40 ml and the total amount of crystalloid administered intravenously to the patient was 200 ml 5% dextrose-0.45% NaCl. Venous blood gases were obtained at the end of the case (Table 1). 10 meq sodium bicarbonate was administered as a slow infusion in the fluid. 130 mg paracetamol was administered intravenously and 50 mg sugammadex was given to reverse the neuromuscular blockade.

The patient was extubated and taken to the PACU for close monitoring. Inhaled adrenaline and cold steam were administered. In our clinic, inhaled adrenaline and cold steam are routinely administered to pediatric patients undergoing head and neck surgery. In our case, their use was specifically indicated due to the patient's tonsillectomy procedure. He was transferred to the ward with stable vital signs. Early pediatric consultation was performed for nutrition. No respiratory distress was observed in postoperative follow-ups. Postoperative analgesia was provided with paracetamol.

Table 1. Intraoperative venous blood gas results.

pH	7.24
pCO_2	28.2 mmHg
pO_2	99 mmHg
sO_2	96.5%
Glucose	169 mg/dL
Lactate	1.7 mmol/L
SBE	-14
HCO_3	11.9 mmol/L

pCO2: Partial Pressure of Carbon Dioxide; pO2: Partial Pressure of Oxygen; sO2: Oxygen Saturation; SBE: Standard Base Excess; HCO3: Bicarbonate.

3. Discussion

The general anaesthetic management without any complications for tonsillectomy of an infant with PA has been described in this case report, with thoroughly monitoring vital signs along with blood gas analysis to track the metabolic state. Anesthesia management in patients diagnosed with propionic acidemia (PA) carries certain risks due to the nature of the disease. The main goal in anesthesia management in these patients is to prevent metabolic decompensation and minimize potential complications of surgical stress [7]. The most important problems that patients with PA may encounter during surgical procedures include metabolic acidosis, hypoglycemia, hyperammonemia and electrolyte imbalances [8]. Therefore, anesthesia management is critical.

In preoperative preparation, the patient's metabolic status should be stabilized prior to surgery. This includes careful monitoring of blood glucose levels, electrolytes, and acid-base balance. In patients on a protein-restricted diet, appropriate dietary adjustments should be made to ensure adequate energy intake in the preoperative period [9]. Fast and short-acting anesthetic agents should be preferred during the induction phase [7]. Propofol should not be preferred as an induction agent because it contains polyunsaturated fatty acids [10]. These agents can reduce metabolic load and provide rapid recovery. Agents such as thiopental and sevoflurane can be used safely in patients with PA; however, the metabolic effects of these agents should be closely monitored. In our case, we used thiopental for induction and sevoflurane for maintenance.

Opioid doses should be carefully adjusted in analgesia management. Short-acting opioids, especially fentanyl, should be preferred [7]. Due to the short duration of the case, no opioids were administered except for 15 mcg fentanyl before ETE (endotracheal intubation). Close perioperative monitoring is important. Blood glucose levels, electrolytes and acid-base balance should be checked frequently during surgery. Constant monitoring is required due to risk of hypoglycemia.

Fluid management of patients should be planned carefully. Hypotension and hypovolemia may lead to increased metabolic stress [11]. In postoperative follow-up, patients should be closely monitored and a nutritional regime should be provided in the early period.

Non-opioid analgesics should be preferred in postoperative pain management [12]. Agents such as paracetamol are safe because their metabolic side effects are minimal.

Given the patient's underlying diagnosis of propionic acidemia, the use of propionic acid derivatives, including ibuprofen, naproxen, and ketoprofen, is generally discouraged due to the potential risk of exacerbating metabolic derangements. These medications can contribute to the accumulation of propionic acid and its toxic metabolites, potentially leading to metabolic decompensation [13]. Therefore, to minimize any metabolic risks, we opted for paracetamol as the primary analgesic agent in this case. Continuous monitoring for signs of metabolic decompensation should be performed and, if necessary, action should be taken in collaboration with a metabolic disease specialist [14].

4. Conclusions

Anesthesia management in patients with PA requires a multidisciplinary approach. The risk of complications can be minimized with close cooperation between the anesthesia team, surgeon, and metabolic specialists. Safely performing the surgical procedure while maintaining metabolic balance forms the basis of successful anesthesia management in this patient group. This case report emphasizes the importance of careful anesthesia application in patients with PA and demonstrates that surgical procedures can be managed successfully.

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Conflict of Interest

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Data Availability

The datasets created and/or analyzed during the current study are not publicly available, but are available from the corresponding author upon reasonable request.

Ethics Approval and Consent to Participate

Written informed consent was obtained from the parents of all infants that were found to be eligible to be included in the study, prior to study enrollment.

Author Contributions

Mehmet Akif Yilmaz: *literature review, visualization, investigation, writing – original draft.*
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