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Case Report

Non-Operating Room Anaesthesia in a Six-Month-Old Female with Propionic Acidemia: Case Report

Eren Demir[1]* and Aytül Taş[2]

[1]*Department of Anaesthesiology and Reanimation, Avcılar Murat Koluk State Hospital, Ministry of Health, Istanbul, Turkey*

[2]*Department of Anesthesiology and Reanimation, Haseki Training and Research Hospital, University of Health Sciences, Istanbul, Turkey*

***Corresponding Author**: Eren Demir, Department of Anaesthesiology and Reanimation, Avcılar Murat Koluk State Hospital, Ministry of Health, Istanbul, Turkey.

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Abstract

Propionic acidemia is a rare metabolic disease. The cases of propionic acidemia can be seen in the operating room, intensive care unit and non-operating room anaesthesia due to the problems related to the complications of the disease. Non-operating room anaesthesia application for magnetic resonance imaging performed in a 6-month-old girl with propionic acidemia is presented with the literature in this article.

Keywords: *Propionic Acidemia; Non-Operating Room Anaesthesia; Deep Sedation; Ambulatory Anaesthesia*

Introduction

Propionic acidemia (PA) is one of inborn metabolic diseases [1]. Clinical findings starting with, vomiting, hypotonia, lethargy, convulsion, acidemia; and can go up to coma [2]. Although it is a rare disease, anesthesiologists may encounter these patients any time. Studies on anaesthesia applications in patients with propionic acidemia are limited in the literature. We aimed to examine the clinical features of patients with propionic acidemia and to emphasize the points that require attention in anaesthesia applications in this case report.

Case Report

A 6-month-old girl with propionic acidemia underwent pre-anaesthetic examination to perform a magnetic resonance imaging (MRI). Cranial MRI was planned because she had convulsions two times about a month ago. She was diagnosed with propionic acidemia at 20-days-old. She had been receiving carnitine and feeding restricted to protein. Physical examination; her weight was 6200 grams, length was 61 cm, head circumference was 42 cm and tone was normal. Breath and heart sounds were normal. Laboratory results were normal. Before the procedure, fingertip blood glucose was 105 mg/dL. Peroral 3 mg midazolam was given as premedication. Electrocardiography (ECG), noninvasive blood pressure (NIBP), peripheral oxygen saturation (SpO_2) and end-tidal carbon dioxide ($ETCO_2$) monitorization were performed. Peripheral vascular access was achieved with 24G branul. Before induction, heart rate (HR) 132/min, NIBP 112/61 mmHg, SpO_2 100% and body temperature was 37°C. Thiopental sodium 2 mg/kg was administered intravenously for induction, and 2 L/min oxygen was given with the mask. Blanket was used to protect body temperature. No additional medication was required. There were no complications during the imaging. After 11 minutes, she was taken to the postanaesthesia care unit (PACU). Fingertip blood glucose was 103 mg/dL in PACU. After an hour follow-up in PACU, the patient was alert and mobile, so she was discharged with recommendations to the parents.

Discussion

Propionic acid is a metabolite formed by the catabolism of isoleucine, valine, threonine, methionine, fatty acid and cholesterol. Propionic acid is converted to methylmalonic acid via propionyl CoA carboxylase, a biotin-dependent mitochondrial enzyme [1]. PA is a metabolic disease that causes propionic acid accumulation due to propionyl CoA carboxylase enzyme deficiency. Early onset PA (the first 6 weeks of life) is starting with vomiting, feeding difficulty, hypotonia, lethargy, metabolic acidosis, and it can progress to coma. Acidosis attacks, encephalopathy, psychomotor retardation, convulsion and dystonia can be seen in patients with late onset (after the first 6 weeks of life) [2]. Protein-restricted diet, biotin and carnitine are used for long-term treatment [3].

The incidence of PA is between 1: 100.000 - 150.000 [1]. Although it is a rare disease; anaesthesiologists may see these patients in the operating room, intensive care unit and non-operating room anaesthesia. The importance of preanaesthetic examination, which is the basis of a problem-free peri- and postanaesthetic period, is even more prominent in patients with propionic acidemia. Mental status, nutritional status, acid-base balance, muscle strength, blood glucose and ammonia levels, and gastrointestinal functions should be examined carefully in addition to routine preanaesthetic examination [4]. Any problems should be corrected in the preanaesthetic period. Because, when anaesthetic drugs and stress factors are added to this status, unwanted complications such as acute attacks may develop. Situations that precipitate metabolic acidosis, such as prolonged fasting, hypoxia, hypotension, dehydration or inappropriate anaesthetic agent should be avoided [5]. The fasting time in the preanaesthetic period should be minimized. Dextrose fluids and sodium bicarbonate can be used, in order to prevent acidosis due to protein degradation during fasting [5]. Hemogram parameters should be examined for anemia and thrombocytopenia. Echocardiography can be performed in the presence of a suspicion for cardiovascular system. Carnitine should be given at the same dose on anaesthesia day. A sample of blood gas should be taken in preanaesthetic period for evaluating the pH, lactate and electrolytes [6].

Non-operating room anaesthesia has some difficulties, such as equipment, environment and procedure. Therefore, more attention should be paid to the non-operating room anaesthesia applications of these patients. We used MR compatible monitor in the presented case. In addition to routine monitoring such as ECG, NIBP, SpO_2, temperature measurement and $ETCO_2$, an arterial catheter can be placed for monitoring blood gas in long-term operations. Fluids with dextrose should be used to prevent hypoglycemia, and fluids such as lactated ringer should be avoided causing lactic acid formation. Gastroesophageal reflux can be seen in patients with propionic acidemia, and, if necessary, rapid-sequence induction should be performed with cricoid compression [4]. Drugs that cause the formation of propionic acid in their metabolites should be avoided. Propofol should not be preferred as an induction agent, because it contains polyunsaturated fatty acids. Muscle relaxants such as atracurium, cisatracurium, mivacurium and succinylcholine metabolised by ester hydrolysis should be avoided [4]. Opioids, inhalation agents and muscle relaxants should be used at the lowest possible doses. Paracetamol may be preferred as postoperative analgesic. Propionic acid derivatives such as ibuprofen, flurbiprofen and naproxen should be avoided [5]. There is no evidence of superiority of an anesthetic agent to another, in patients with propionic acidemia. The choice of dosage and medication should be based on the anaesthetist experience and the clinical status of the patient. Because of the increased risk of systemic toxicity of local anaesthetics, regional anaesthesia is controversial in these patients who have mitochondrial dysfunction. However, it can be applied in selected cases, Soberon., *et al.* reported successful and problem-free peripheral nerve block application to the patient with propionic acidemia [7]. Blood glucose monitoring should be performed during the perianesthetic period. Hypoventilation, hypoxemia, dehydration and hypothermia, which may worsen the patient's clinical status, should be avoided. More attention should be paid to sterilization in neutropenic patients [4].

Patients should be followed up in PACU until they return to their basal status and should be taken care with regard to complications such as hypoglycemia and respiratory distress. PACU follow-up is becoming more important in non-operating room anaesthesia, because of the patients' going home directly. Before discharge, patients should be fully recovered, and detailed information should be given to the family. The patient should be fed as soon as possible, for preventing hypoglycemia.

Conclusion

Non-operating room anaesthesia has various risks. The risk increases even more in pediatric patients swith diseases such as PA. Careful preanesthetic examination and postanesthetic care mayprevent the development of these problems. Studies on non-operating anaesthesia applicationsin pediatric patients with metabolic disease such as propionic acidemia are limited. We believe that it will be beneficial to do more studies on this subject.

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