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Case Study and Anesthetic Strategy of an Infant with Propionic Acidemia That Undergone a Cardiac Surgery

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Abstract

According to literature reviews, the paucity of infants with propionic academia was manifested. There are a variety of important medical issues associated with this genetic abnormality. Although there are an enormous range of methods and techniques to identify and treat metabolic defects, the requisite of an anesthesiologist for a specific plan, yet is not comprehensively answered. In this case, we have presented a 17-month old boy with propionic academia who was also suffered from frequent respiratory infection, tachypnea and failure to thrive; our echocardiography demonstrated both atrial septal defect (ASD) and patent ductus arteriosus (PDA). He was candidated for required closure surgery. In this article we provided information regarding propionic acidemia and its perioperative anesthetic implication especially in a cardiac surgery.

Keywords

Anaesthetic Techniques, Propionic Academia, ASD, Cardiac Surgery

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1. Introduction

Propionic acidemia is a congenital disease caused by an autosomal mutation. Propionic acid can be produced in two different way: Or there is a deficiency in propionyl coenzyme carboxylase or there are some course of odd chain fatty acid fermentation in the alimentary tract of these patients. Either of the two situation would lead to some defects in amino acid and fatty acid metabolism[1]. Propionic acid inhibits citric acid cycle enzymes and develops ketoacidosis [2, 3].

Along with the acidosis, manifestations of the disease may include developmental retardation, seizures, Coma, hypotonia, episodic vomiting, gastroesophageal reflux, protein intolerance, hypogammaglobulinemia, hyper ammonemia (caused by inhibition of acetyl glutamate synthetase by propionic acid), osteopenia, bone marrow dysfunction, cardiomyopathy, and pancreatitis [4].

2. Case Report

A 17- month old boy with propionic acidemia was scheduled for elective ASD and PDA closure surgery. He was a term child born to a family with no history of genetic diseases. Initial diagnosis was made at 4 days of age when he presented with lethargy and hyperammonemia. He was treated with L-arginine, L-carnitine, folic acid and B6, B12 Vitamins; also captopril, Laxis and Lanoxin was administered for underlying cardiac disease. Prior to his admission, his hyperammonemia and acidosis was corrected.

Physical examination revealed a 8.3 kg male with a blood pressure of 105/70 mmHg, a Heart rate of 100 bpm, and a respiratory rate of 20 breaths/min. Auscultation of the heart showed:S1 OK-S2 single- 3/6 Smm at Left Sternal Boarder (LSB) . In lung examination breathing sounds was normal. Further, the chest radiography demonstrated increased

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interstitial markings and cardiomegaly.

Laboratory results were as follows: Ammonia (plasma) 0.54 μg/ml, Lactate (plasma) 17 mg/dl, Hb 10.6 gr/dl, Alkaline phosphatase normal, a platelet count of 243,000, Na 149 meq/lit, K 5.1 meq/lit, PHa 7.46, $PaCO_2$ 29.1 mmHg, PaO_2 69.7mmHg, HCO_3 24.2, Tco_2 21.1, $SpaO_2$ 95.2%, BE -0.5, BB 47.5, O_2 Content 19.3; Urinalysis and coagulation studies were normal.

Transthoracic echocardiography demonstrated ASD2nd(secondum type ASD), Small PDA and dilated main pulmonary artery.

Angiography demonstrated large ASD2, very small PDA and Top normal pulmonary artery pressure.

In the operating room, routine monitors were used, including noninvasive blood pressure, electrocardiography, pulse-oximetry; invasive blood pressure (left radial artery), Central venous pressure (Left subclavian vein) and esophageal temperature was also established after induction.

Induction of anesthesia was initiated with sevoflurane by mask, pancuronium 1.2 mg and fentanyl 150 μg. After six minutes adequate ventilation through face mask, the patient was tracheally intubated without trauma on the first attempt with a 4.5 uncuffed endotracheal tube, and the tube was taped at 12 cm at the lips.

Pancuronium 0.5 mg in the beginning of cardiopulmonary bypass (CPB), Fentanyl 0.1μg/kg/min and O_2 were used for anesthetic maintenance with frequent ABG analysis and blood sugar(for prevention of hypoglycemia) measurement throughout the case.

The maintenance fluid was Dextrose %5 (D5) 25 ml/h and NS 25ml/h through a continuous infusion pump.

Following sternotomy surgical hemostasis was obtained. About ten minutes prior to routine aortic and right atrial cannulation Heparin 3000 units was administered as a bolus. Anticoagulation was monitored using a Medtronic II activated clotting time (ACT) device with celite based ACT tubes and a Hemochron device. The ACT rised to level of 653 sec. When adequate anticoagulation established, CPB was initiated in the usual fashion and conducted under moderate hypothermia (30°C). After institution of CPB, cardiac arrest was achieved by antegrade cardioplegia. The ASD was closed with pericardial patch and PDA was also ligated. Rewarming was commenced and the patient successfully weaned off CPB. The total CPB time was 41 min, the patient was decannulated and all surgical sources of bleeding stopped.

Total priming volume was 760cc which included 50cc of packed cell.

Total urine output during operation was 40 ml.

The blood glucose, lactate, Hematocrit, Hb, Ca, Cl, K, Na and blood gasses was measured (Table 1)

Table 1. Blood glucose, lactate, hematocrit, Hb, Ca, K, Na and Blood gasses results

	PH	PO2	Sao2	PCo2	HCo3	TCo2	BE	Na	K	Hb	HT (%)	BS	Lactate	Cl	Ca
Before CPB FIO2=1	7.42	343	100%	36	23	24	0	135	3.5	9.6	31	235	1.1	104	1.4
After institution of CPB	7.39	284	100%	42	25	26	+1	136	4.4	7.1	23	114	3.3	111	2.2
After rewarming was commenced	7.42	450	100%	34	22	23	-1	133	3.7	6.8	22	171	2.1	108	1.6
After weaned off CPB	7.36	379	100%	39	22	23	-2	137	4.1	10.5	34	112	2.5	104	1.2

Table 2. Laboratory results

	Before surgery	ICU Admission	4 hours Later	8 hours later	2nd day	3rd day
BS	125	172	248	127	172	120
BUN	13	11	19	-	20	14
Creatinine	0.4	0.5	0.5	-	0.5	0.5
AST	61	-	119	-	-	76
ALT	50	-	22	-	-	24
Ammonia (plasma)		-	-	-	1.1	0.55
Lactate (plasma)		-	-	-	29	27

Neutralizing the effect of heparin, protamine 40 mg was slowly infused at the end of CPB. Packed cells 60 ml was infused for correction of low Hb at the end of CPB. He was transferred to the intensive care unit under continuous monitoring where his lungs remained mechanically ventilated. The patient was transferred to the operating room 3.5 hours later due to 800 ml bloody drainage. Induction of anesthesia was initiated with Fentanyl 100 mg, Pancuronium 1 mg. Isoflurane 0.4% and O_2 were used for anesthetic maintenance. Following sternal Exploration a source of active bleeding on Right Atrium was stopped by suturing, the chest was closed and the patient was transferred to the intensive care unit where

his lungs remained mechanically ventilated, because of weak respiratory effort; During 16 hours, he gradually developed good respiratory effort, and reflexes. At last, he was successfully extubated 2 hours later and the remainder of his ICU stay was uneventful and serial blood gas measurements was acceptable (Table 2).

3. Discussion

The risk of an acidosis crisis in Congenital cyanotic heart lesion is more in lesions with extracardiac or intracardiac left-to-right shunts and increased pulmonary blood flow that can protect the patient from hypoxemia, the primary concern in the patients is to avoid hypotension that precipitates inadequate tissue perfusion and acidosis.

When patients with propionic acidemia are fasting, they require glucose in their IV fluids to suppress protein catabolism and subsequent acidosis. Dextrose therapy may be needed.

Propionic acid produced by protein breakdown initiated by fasting or surgical stress. Bicarbonate, at the patient's usual daily dose, is used to limit this effect. Lactic acid-containing fluids (e.g., Lactated Ringer's) should be avoided because contribute to the patient's acid load. Glucose level, PH, and ammonia should be included in the preoperative laboratory tests.

Intraoperatively, a rapid sequence induction should be considered in patients who have history of reflux or vomiting, also cricoid pressure application is advised. Muscle relaxants metabolized by ester hydrolysis (succinylcholine [5], cisatracurium [6], atracurium [7], and mivacurium [8]) should be avoided because their metabolites include odd-chain organic molecules. Propofol is banned because it is an aqueous solution containing soybean oil, which consist of high polyunsaturated fats [9], that may be metabolized to propionic acid [10]. Ibuprofen, Ketoprofen, Naproxen, and oxaprozin [11] are also forbidden because they are derived from propionic acid.

Blood glucose level should be measured intermittently and if prolonged anesthesia is predicted, arterial PH should be monitored. Probable lactic acidosis during CPB can be prevented by maintaining tissue perfusion with adequate pump flow rate. During this period, flow rates should be chosen more cautiously to provide systemic oxygen delivery.

Although, routinely utilization of systemic hypothermia during pediatric CPB with several operations was accepted among practitioners, it should be considered that severe hypothermia can reduce tissue perfusion that foster acidosis. Our recommendation within this period is using of full flow rates and avoidance of severe hypothermia.

These patients may be particularly sensitive to the central nervous system depressant effects of volatile anesthetics and narcotics analgesics [12].

Sterile technique should be carefully maintained because infection precipitates acidosis.

Recent improvement in the dietary management of these patients has made osteoporosis less likely to occur, and fractures are a rare problem [13]. Similarly, the cardiomyopathy is rarely seen because of the use of L-Carnitine, which promotes propionic acid excretion from the body.

Postoperatively, patients with upper airway obstruction or fatigue may be prone to develop respiratory distress. The use of humidified supplemental oxygen may be helpful.

Post-operative hypothermia might occur, which results in shivering, high oxygen consumption, and increased cardiac stress. The associated increased CO_2 production may cause respiratory acidosis; moreover, inadequate tissue oxygen delivery may cause metabolic acidosis. Thus, suitable temperature conditioning should be applied.

4. Conclusion

In conclusion, it should be considered that there are challenging circumstances facing the whole perioperative period. Though, the basic concepts of managing these patients are discussed recently, different approaches might be chosen by different anesthesiologists. We hope further studies would lead to a thorough strategy for patient's management. By the way, the following are a brief review of the author's suggestions:

Preoperative evaluation of the patient with propionic acidemia should focus on nutritional state, mental status, muscle tone, acid-base balance, and gastrointestinal function. These factors have the greatest impact on anesthetic management. The primary concern is to avoid events that leads to metabolic acidosis. Acidosis can be initiated by inadequate caloric intake, hypoxia, hypotension, hypothermia, dehydration, or the use of an inappropriate anesthetic.

In summary, anesthetic managements should be designed to avoid metabolic acidosis and prevent airway complications. Acidosis can be prevented by maintaining adequate tissue perfusion, avoidance of hypothermia and Hypoxemia; Protein catabolism can be prevented by providing dextrose in IV fluids.

Tracheal extubation is delayed until the patient has regained baseline muscle strength. Post-operative vigilance is essential even if extubation is delayed, because upper airway obstruction and respiratory fatigue can produce respiratory distress.

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