Anaesthetic Management of a Child with Propionic Acidemia

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ABSTRACT

Propionic acidemia (PA) is an inherited autosomal recessive disorder of metabolism caused by a deficiency of propionyl CoA carboxylase, an enzyme that catalyses the conversion of propionyl CoA (PCA) to methylmalonyl CoA, inside the mitochondria, leading to inadequate metabolism of propionyl CoA causing hyperammonemia and metabolic acidosis. Children with PA require dextrose infusion to avoid protein catabolism. This child presented with severe metabolic decompensation and required urgent venous cutdown as there was a failure in establishing a peripheral intravenous line.

Key Words: *Propionic acidemia, Propionyl CoA, Emergency, Mitochondrial disorder, Organic acidemias.*

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INTRODUCTION

Propionic acidemia (PA) is an inherited autosomal recessive disorder of metabolism caused by the deficiency of propionyl CoA carboxylase, an enzyme that catalyses the conversion of propionyl CoA (PCA) to methylmalonyl CoA inside the mitochondria.[1] Limited literature is available regarding the anaesthetic management of such cases, especially in an emergency situation.

The rationale of writing this case report was that children with PA often require cannulation and frequent dextrose infusion in order to prevent or treat metabolic crisises. Provision of anaesthesia in this emergency situation without any intravenous access and the possibility of airway and cardiovascular collapse during induction was a challenge in this case. We have demonstrated the use of an interosseous needle for the induction of anaesthesia in emergency situations.

CASE REPORT

Written informed consent was obtained from the parents before writing the case report. A 13-month boy, weight 8.5 kg, required urgent anaesthesia for venous cutdown. The child was born at term and delivered vaginally. His family history included the death of all four older siblings either as neonates or in infancy. His milestones were delayed, and he had a past medical history of seizures two months back, which were self-aborted.

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The child initially presented to the paediatric clinic at six months of age. His laboratory investigations showed urinary ketones of 3+ and hypoglycemia. He was immediately treated with 25% dextrose and L-carnitine. He was diagnosed as suffering from PA based on the history of repeated chest infections and recurrent hypoglycemia. He visited the hospital multiple times due to decompensated metabolic acidosis and ketonuria. He was put on specialised formula milk (Similac 1 and Anamix infant) and a strict diet plan that included vegetables, fruits, and rice but no meat.

During the current visit, the child presented with severe metabolic decompensation and required urgent venous access for administration of dextrose infusion after failure in establishing a peripheral line. He was rushed to the operating room as an emergency. Preoperative parameters were a pulse rate of 137 beats /min with normal sinus rhythm, blood pressure of 80/40 mmHg, SpO_2 of 97% on room air, and a respiratory rate of 25 breaths/min. On physical examination, he was a thin, lean child, drowsy, and lying in his mother's lap. Other cardiovascular and respiratory examinations were normal. He had taken oral replacement therapy by mouth five hours back.

After the application of standard American Society of Anaesthesiology (ASA) recommended monitors, that is, ECG, noninvasive blood pressure, temperature, respiratory rate, and SpO_2, anaesthesia was induced with 100% O_2 and sevoflurane, 2-4%. An interosseous line was placed by the surgeon using a Jamshidi needle of 15/15 G in the right tibia.

After the establishment of an interosseous access, a size 1.5 laryngeal mask airway (LMA) was placed, maintaining spontaneous ventilation and anaesthesia was continued with sevoflurane 2% in N_2O/O_2 50:50% mixture. The child desaturated to 93% on FiO_2 of 50%, this required FiO to be intermittently increased to 100%.

A venous cutdown was performed in the left saphenous vein, and a 4 Fr. central venous catheter (CVC) was inserted. Free flow of blood was confirmed, and 0.45% normal saline and 5 % dextrose

infusion was started. The patient remained hemodynamically stable. Arterial blood gas was done intraoperatively after insertion of CVP, which showed a pH 7.30, $PaCO_2$ 37.8 mmHg, and a HCO_3 of 18.5 meq/dl.

After completion of the procedure, the child was allowed to wake up, and LMA was removed. He was shifted to the post anaesthesia care unit. The child remained in special care for 24 hours and then shifted to the ward and discharged home after two days of hospital stay.

DISCUSSION

PA is a rare autosomal recessive disorder characterised by a deficiency of propionyl CoA carboxylase, which along with biotin, is crucial for the conversion of propionic acid to D-methylmalonyl CoA.[2]

PA has an incidence of 1 in 100,000-350,000 in United States and is found more often in the Inuit population of Greenland, some Amish communities, and Saudi Arabians.[2]

No data is available on the incidence of PA in Pakistan, however, Cheema *et al*. demonstrated a frequency of 18 out of 180 patients with organic acid disorders.[3]

The challenges that we faced in the management of this child were:

Firstly, this was an emergency situation as children with PA needed intravenous glucose to reduce protein catabolism, and acidosis, and the case could not be delayed.

Secondly, inhalational induction was done with sevoflurane in this child to avoid propofol, which contains polyunsaturated fats. Small amounts of fat present in propofol is converted into propionic acid in the body.

In addition, children with PA have generalised muscular hypotonia which can lead to complete airway collapse on induction with an intravenous agent; therefore, vigilance is required at induction even if an intravenous line is in place. Inhalational induction offers a safety mechanism whereby anaesthesia can be lightened if airway obstruction occurs.

Thirdly, intravenous access was a big challenge in this case. So, we managed the child by placing an interosseous needle soon after the induction of anaesthesia in view of the anticipation of cardiac decompensation and untoward effects secondary to inhalational anaesthesia.

Fourthly, extubation is again a crucial period in these patients. Harker *et al*. demonstrated respiratory depression in a child suffering from PA after extubation is secondary to mucus plugs and secretions.[4] We extubated the child once he was fully awake, in order to avoid aspiration of secretions.

Limited literature is available regarding anaesthetic management of these children. Kim *et al*., reported successful liver transplantation in a child with PA.[5] Young *et al*. have reported a case in which the child expired after liver transplantation secondary to liver failure and severe metabolic acidosis.[6]

To the best of our knowledge, our search did not reveal any case report where an interosseous needle has been used in the management of these patients. We, therefore, recommend the use of an interosseous needle where peripheral intravenous access is not possible.

PATIENT'S CONSENT:
Written informed consent was obtained from parents of the patient to publish this case.

COMPETING INTEREST:
The authors declared no competing interest.

AUTHORS' CONTRIBUTION:
SS: Helped in preoperative preparation, perioperative care, consent from the parent, literature search and initial write-up of case summary, introduction, report and discussion.
FAK: Helped in supervision during the clinical care, proof reading, and editing of the manuscript.
All the authors have approved the final version of the manuscript to be published.

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