



Anesthetic management of a child with methylmalonic acidemia for lens aspiration – a case report

Anwar ul Huda, Mohammad Yasir

ABSTRACT

Security Forces Hospital,
Salah Ad Din Al Ayyubi Road,
Ad Dhubbat, Riyadh 12625,
(KSA)

Correspondence: Dr Anwar ul
Huda, Security Forces
Hospital, Salah Ad Din Al
Ayyubi Road, Ad Dhubbat,
Riyadh 12625, (KSA)
Phone: +966502654719
E-mail:

hudaanwar90@yahoo.com

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We report a case of a child with methylmalonic acidemia who presented to our hospital for lens aspiration surgery. Methylmalonic acidemia is an inherited disorder of protein catabolism. Its clinical effects can vary from mild to severe, and include lethargy, vomiting, hypotonia, hypothermia, failure to thrive and encephalopathy. The main concern in treatment of these patients is prevention of acidemia by identifying precipitating factors. It is especially important to acutely manage fluids and electrolytes before, during and after surgery.

Key words: methylmalonic acidemia; methylmalonic acid; Acidosis/genetics

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INTRODUCTION

Methylmalonic acidemia (MMA) is an inborn error of metabolism with an incidence of estimated 1 in 50,000 people.¹ It is an autosomal recessive disorder and is characterized by partial or complete deficiency of methylmalonyl-CoA mutase enzyme.² Patients with this disorder have enhanced protein catabolism resulting in severe metabolic acidosis, hyperammonemia and ketosis.¹ Those with complete deficiency of enzyme usually present within first few days to weeks of life with sudden deterioration of general condition and acidemia progressing to coma and death. Cases with delayed onset can present at any age with more heterogeneous clinical features.¹ Overall, the outcome of this disease remains poor despite effective treatment with a low protein diet and carnitine except for vitamin B12-responsive forms of MMA, which carry a better outcome if diagnosed and treated in a timely manner.³ We describe a case report of successful anesthesia management of a 3 year

old child with MMA undergoing ocular lens aspiration surgery.

CASE REPORT

A 3 year old male child, weighing 11.2 kg, was scheduled for elective lens aspiration surgery for congenital cataract. He was diagnosed with MMA at the age of 9 months, when he was admitted with vomiting, diarrhea, and cough. He was positive for organic acid on urine organic acid qualitative test, which was later confirmed on quantitative analysis. He had history of developmental delay in achieving milestones. Parents were advised to give the child low protein diet and Polycitra- K to decrease acidity.

Patient was admitted a day before surgery and was started on 10 % dextrose water at 45 ml/hr as prescribed by pediatrician to prevent hypoglycemia. His preoperative blood investigations were within normal range, including serum bicarbonate of 22 mmol/L (18-26 mmol/L). On the day of surgery, before moving the patient to operating room, he

received intravenous (IV) midazolam 1 mg to allay anxiety. In the operating room, ASA standard monitors were applied. Intravenous dextrose 10% was continued. Preoxygenation was started and anesthesia was induced with propofol 30 mg and fentanyl 10 mg IV, without the use of any muscle relaxant. Endotracheal intubation was done with size 4.5 mm RAE micro-cuffed endotracheal tube. Anesthesia was maintained with sevoflurane 0.8-1.0 MAC in oxygen air mixture and inj. remifentanyl 0.03 - 0.07 mg/kg/min. Intraoperative temperature was controlled with forced air warming device. Inj. paracetamol 250 mg was administered intraoperatively. The surgery lasted for 1 hour and was uneventful. At the end of the operation, remifentanyl infusion was stopped and inhalational agents were discontinued. Additionally, fentanyl 5 mg was given before extubation to control postoperative pain. Patient was extubated once he met the criteria. Postoperatively, he remained stable and oral feeds were started 4 h after surgery. Patient remained admitted for one day and then was discharged home.

DISCUSSION

Patients with MMA have increased protein catabolism, preoperative fasting needs to be limited as it can increase protein catabolism even more. Additionally, the stress response of surgery increases the process of catabolism. Anesthesia itself may affect amino acid metabolism resulting in increased levels of

MMA; for example nitrous oxide influences metabolism of Vitamin B12. However, the major concern in MMA patients is acute metabolic failure.⁴

In our case, we tried to address these concerns. We limited preoperative fasting to 6 h and IV 10 % dextrose was started preoperatively in order to minimize hypovolemia and protein catabolism. Perioperatively, every measure was taken to prevent hypoxia, hypercarbia, hypothermia, hypoglycemia and any such stress that can precipitate acidosis. IV midazolam was used to reduce preoperative anxiety in the child.

Ktena et al. in their study concluded that it is safe to administer propofol and atracurium in patients with MMA.⁵ Also, postoperative nausea and vomiting (PONV) prophylaxis is necessary in these patients as they are prone to nausea and vomiting and subsequent dehydration results in acidosis.² The authors did not find any literature regarding use of remifentanyl in MMA patients but it was successfully used in our case without any adverse reactions.

In conclusion, patients with MMA can potentially develop severe complications and anesthesia management need to be tailored very carefully.

Conflict of interest: None declared by the authors

Authors' contribution:

AUH – Concept, conduct of case, writing and editing the manuscript

MY - Conduct of case, writing and editing the manuscript

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