

Ketamine and dexmedetomidine sedation for brain magnetic resonance imaging in methylmalonic acidemia

Methylmalonic acidemia (MMA) occurs due to the deficiency of methylmalonyl-CoA mutase, which converts methylmalonyl coenzyme A (CoA) to succinyl-CoA, leading to the accumulation of methylmalonic acid.^[1] There is also an associated deficiency of propionyl-CoA carboxylase resulting in the accumulation of propionic acid. This leads to a state of acute metabolic decompensation with acidemia and hyperammonemia, and clinically, patients present with vomiting, seizures, developmental delay, mental retardation and renal failure.^[1] Anaesthetic challenges include avoiding excessive fasting, which can initiate protein catabolism leading to metabolic decompensation, adequate depth of sedation, maintenance of euvolaemia and providing enough energy to meet the metabolic demand. Apart from this, careful selection of anaesthetic drugs is important to have an uneventful outcome.^[2] We present a 1-year, 8-month-old female child, weighing 7.8 kg, diagnosed with MMA. She received sedation with a combination of ketamine and dexmedetomidine (keta-dex) for magnetic resonance imaging (MRI) of the brain for the evaluation of global developmental delay. Previously, she was admitted with hyperammonaemia, hyperlactatemia and high anion gap metabolic acidosis and was metabolically optimised with a low protein diet, l-carnitine, hydroxycobalamin, lactulose, sodium benzoate, biotin and sodium bicarbonate therapy.

On the day of MRI, nil per orally for 2 hours for clear liquids and 6 hours for solids was advised. In the patient holding area, intravenous (IV) access was secured with prior application of prilocaine and lignocaine cream to avoid pain and stress. In the MRI suite, after attachment of standard American Society of Anesthesiologists monitors, she was sedated with IV ketamine 1 mg/kg and IV dexmedetomidine 1 µg/kg over 10 minutes, followed by maintenance with IV infusion of dexmedetomidine at 0.5 µg/kg/hour to maintain a score of 2 as per the Observer's Assessment of Alertness/Sedation Scale (OAA/S).^[3] Oxygen supplementation was given via a face mask. Due to an isolated patient movement, dexmedetomidine infusion was increased to 1 µg/kg/hour, and propofol 10 mg was

given. The duration of MRI was 40 minutes, following which the child was observed in the post-anaesthesia care unit and received 5% dextrose in normal saline at 4 ml/kg over an hour as maintenance fluid.

In this case, we used intravenous sedation with 'keta-dex' with the patient on spontaneous respiration and end-tidal carbon dioxide monitoring. We used 'keta-dex' because of the previously reported association of metabolic decompensation with propofol infusion for organic acidemia.^[2,4-5] Secondly, as a combination, both complement each other and provide adequate sedation and stable haemodynamics.^[6] Nevertheless, propofol is safe for both the induction and maintenance of anaesthesia/sedation in metabolically stable patients of MMA,^[4] such as this patient, who was metabolically optimised before sedation. Safe use of barbiturates and sevoflurane has also been described.^[2] Nitrous oxide and propionic acid-derived analgesics such as ibuprofen are not recommended.^[2] Along with the maintenance of euvolemia, 10% glucose at 7–8 mg/kg/minute supplementation is recommended for a short course of time to prevent protein catabolism and meet the energy demand.^[1] The management should be closely monitored with arterial blood gas (ABG) analysis wherever possible. Considering the short duration of sedation, we did not monitor ABGs.

Avoidance of prolonged fasting, hypovolemia, selection of anaesthetics and metabolic optimisation are crucial while managing patients of MMA. 'Keta-dex' combination is a reasonable option for sedation in these patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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