Letters to Editor

Bypassing conventional anaesthetics: Dexmedetomidine sedation in MEGDEL syndrome

Sir,

3-Methylglutaconic aciduria (MEG) with sensori-neural deafness (D), encephalopathy (E), and neuroradiological evidence of Leigh-like disease (L) (MEGDEL) syndrome is one of the rare mitochondrial disorders with a prevalence of 0.09:100,000. Clinical features include progressive myopathy, hypoglycaemia, feeding difficulty, developmental delay, failure to thrive, acute liver failure, arrhythmias and cardiac failure.[1]

We report a case of a 1-year-old female child, weighing 8 kg, scheduled for magnetic resonance imaging (MRI) and computerised tomography (CT) under anaesthesia. The child was diagnosed to have MEGDEL syndrome on the basis of genetic and biochemical testing in the neonatal period. She presented with global developmental delay, bilateral profound hearing loss and bilateral pyramidal signs. There was no history of seizure disorder.

A careful anaesthesia plan was chalked out considering the concerns of using conventional anaesthetic agents like propofol and sevoflurane for sedation. Since both these agents are contraindicated in a child with mitochondrial dysfunction, we planned to use dexmedetomidine infusion to maintain sedation during the MRI procedure. The child was admitted on the morning of the procedure and was kept fasting as per American Society of Anesthesiologists guidelines. Intravenous (IV) maintenance fluid with dextrose normal saline at the rate of 25 ml/hour was started 2 hours prior to the procedure and administered till the induction of anaesthesia. Pre-induction blood glucose level was 70 mg/dl and a bolus of 15 ml of dextrose normal saline was also administered prior to induction. Sedation was induced with IV ketamine 0.25 mg/kg, IV dexmedetomidine 0.5 μg/kg and IV midazolam 0.1 mg/kg, in titrated bolus doses. Maintenance of sedation was done with IV dexmedetomidine infusion at the rate of 0.5 μg/kg/hour. The baseline core temperature was 36.4°C and the child was carefully wrapped in warm clothes prior to shifting inside the MR gantry. The imaging period was uneventful and was completed in 35 minutes. The child was next shifted to the CT room with ongoing sedation and the scan was completed in 4 minutes duration. The vital signs of heart rate, respiratory rate, and saturation were continuously monitored and were within normal limits. Following the completion of MRI and CT, dexmedetomidine infusion was discontinued, and the child was monitored for awakening, vital signs, temperature and glucose. The child woke up after 15 minutes and vital signs remained stable during the recovery period. Post-procedure period in the ward was uneventful and the child was discharged the next day.

In this report, we want to highlight the use of dexmedetomidine in a patient with MEGDEL syndrome. Dexmedetomidine was used instead of propofol or sevoflurane based anaesthesia, to avoid the development of malignant hyperthermia due to inhaled anaesthetics or mitochondrial dysfunction due to propofol infusion and inhaled anaesthetics. The reduced ability to generate adenosine triphosphate and use oxygen can result in these patients being at more risk of developing rhabdomyolysis and anaesthesia induced malignant hyperthermia. Propofol, though considered safe in malignant hyperthermia, has the same risk as inhaled anaesthetics in causing mitochondrial dysfunction.[2]

Centrally acting alpha-adrenergic agonists like dexmedetomidine can be used for sedation as an alternative to propofol. Dexmedetomidine does not depress respiration, protects airway reflexes, provides good anxiolysis and sedation.[3] It has been reported to be safe in patients with mitochondrial disease since the mechanism of action in the central nervous system differs from commonly used general anaesthetics. Successful trials have been conducted using non-triggering anaesthesia with dexmedetomidine in patients with mitochondrial disease.[4,5]

We also made efforts to reduce the metabolic burden of the patient which includes avoidance of prolonged fasting and hypoglycaemia, prevention of hypovolaemia and maintenance of normothermia.[6]

In remote location anaesthesia like an MRI suite, dexmedetomidine is a safer alternative to propofol or sevoflurane for children with mitochondrial dysfunction undergoing sedation. In addition, strategies to decrease the metabolic burden in the perioperative period should be stringently employed. Post-anaesthesia monitoring in a hospital setting is prudent for the first 24 hours.

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Conflicts of interest

There are no conflicts of interest.

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