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LETTER TO THE EDITOR

Ketamine–midazolam anesthesia for an infant with arthrogryposis multiplex congenita: a case with decreased myocardial contractility

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To the Editor:

Arthrogryposis multiplex congenita (AMC) is mainly characterized by multiple joint contractures and is often accompanied by other systemic anomalies, including cardiovascular abnormalities [1]. Anesthetic management of AMC patients carries risks of difficult airway, difficult IV access, and a malignant hyperthermia (MH) episode.

We here present an infant with AMC and with decreased myocardial contractility in whom we used ketamine–midazolam for anesthetic management.

A three-and-a-half-month-old, 2840-g preterm female infant who had been receiving mechanical ventilation in a neonatal intensive care unit since birth with the diagnosis of neonatal respiratory distress syndrome was scheduled for bilateral keratitis because of possible risk of corneal perforation.

The birth history revealed that the infant was born by Caesarean section at 33 weeks of gestation. The indication for caesarean section was initiation of preterm labor and polyhydramnios.

On admission to our hospital, she also had neonatal bacterial septicemia, AMC, and keratitis. On examination, flexion contractures of the hands and feet were noted (Fig. 1). She had an orotracheal tube and a nasogastric feeding tube. Her medication included dopamine, dobutamine, and furosemide for depressed myocardial contractility, and piperacillin–tazobactam and vankomycin for septicemia. Cranial ultrasonography detected bilateral periventricular-intraventricular hemorrhage.

In the operation room, monitoring included ECG, SpO_2, $PETCO_2$, NIBP, and axillary temperature probe. Her blood pressure was 76/35 mmHg, heart rate was 142/min, and SpO_2 was 97. After 0.02 mg/kg atropine intravenously, 0.06 mg/kg midazolam and 1.5 mg/kg ketamine were given. Anesthesia was maintained with a 50% O_2/50% N_2O mixture and no additional anesthetics were needed during amniotic membrane transplantation operation for left corneal ulcer. Duration of operation was 25 min, and anesthesia was uneventful. Tachycardia, a rise in $ETCO_2$, or temperature was not seen during surgery.

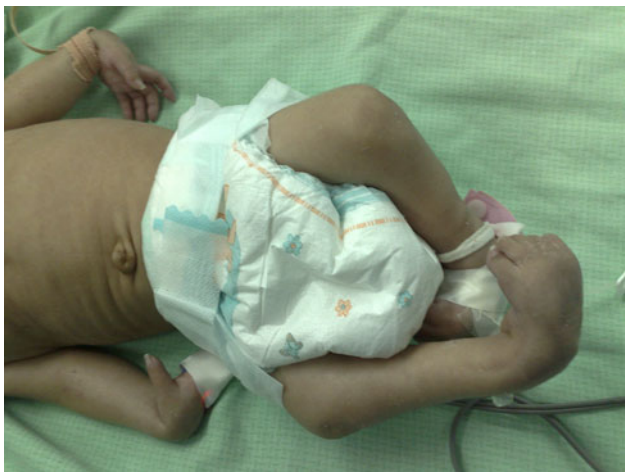

Fig. 1 Flexion contractures of the infant with AMC

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Patients with AMC are at increased risk of morbidity and mortality from anesthesia, and one of the major concerns of anesthetic management is the possible risk of MH. It has been reported that inhalational anesthetic drugs, which carry risk for patients with decreased myocardial contractility, may trigger MH reactions in AMC patients [2]. Apart from MH, AMC patients may have a hypermetabolic response unrelated to anesthetics. Increases in fever, $ETCO_2$, and heart rate occur in cases with AMC [3].

So, avoiding the use of volatile anesthetics minimizes both the risk of malignant hyperthermia/hypermetabolic response and the possibility of anesthetic-induced myocardial depression, because all inhalational anesthetics have this effect in a dose-dependent manner.

The choice of ketamine and midazolam helped to provide stable hemodynamics in our infant with decreased myocardial contractility. Ketamine also is a good choice for children who is at risk of the development of malignant hyperthermia [4]. Although there is some controversy regarding the use of ketamine in these patients, work by Dershwitz et al. [5] has suggested MH did not develop in 76 MH-susceptible swine in which ketamine was used as an induction agent.

In conclusion, the use of ketamine–midazolam resulted in stable hemodynamics and avoidance of use of volatile anesthetics which has a potential for myocardial depression.

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