[NM-160] Ventricular Fibrillation on Induction of Anesthesia with Sevoflurane in a Patient with Prolonged QT Syndrome.

Arnold M, Istaphanous G
Childrens Hospital Los Angeles , Los Angeles , CA, USA

Introduction: Timothy syndrome, also known as long QT syndrome type 8, is a rare multisystem autosomal dominant disorder characterized by syndactyly, dysmorphic features, cognitive deficits, congenital heart defects and arrhythmias. This condition is extremely rare with only 25 molecularly confirmed cases reported worldwide. It is caused by a mutation in the CAv1.2 L-type calcium channel gene. Identification of the common mutation p.Gly406Arg in exon 8A of CACNA1C confirms the diagnosis.(1,2) Inhalational anesthetics such as sevoflurane are known to prolong QTc interval. However, the clinical significance is unclear and it is controversial whether it should be avoided in patients with prolonged QT.(3,4)
Case Description: A full term 6-month-old 6.2kg female with a history of Timothy syndrome presented for surgery for an epicardial automatic implantable cardioverter-defibrillator. At 23 weeks of gestation she was diagnosed with 2:1 Atrioventricular heart block and was eventually found to have 2-3 toe syndactyly and a left eye coloboma. This prompted an EKG at birth, which showed 2:1 AV block and prolonged QTc. She was monitored in the NICU for 3 weeks, had no ventricular dysrhythmias and was discharged home on propranolol. Baseline EKG one week prior to surgery revealed sinus bradycardia at a rate of 107BPM and a prolonged QTc of 583. Echocardiography was otherwise normal. On the day of surgery, she was orally pre-medicated with 0.75mg/kg midazolam. Induction of anesthesia with sevoflurane and oxygen took place following the placement of a pulse oximeter, blood pressure cuff, EKG and pacer pads. Ventricular fibrillation and hypotension were noted within thirty seconds of induction. Chest compressions were started immediately and she was successfully defibrillated with 10 Joules. She instantaneously recovered and an EKG revealed a 2:1 AV block with a rate in the 100’s, at which point she was treated with magnesium and sevoflurane was discontinued. Maintenance of anesthesia was achieved with propofol, fentanyl and midazolam with return to baseline QTc and sinus rhythm within 10 minutes. The surgery was completed with no other events and she was transported to the cardiac ICU for postoperative observation.
Discussion: Whether all patients with congenital prolonged QT should avoid exposure to volatile anesthetics is controversial. However patients with Timothy syndrome may be dangerously susceptible to their arrthymogenic effects and extra measures, such as availability of anti-arrhythmic medications and defibrillator pads should be considered.

References:
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