Hyperkalemic cardiac arrest following cardiopulmonary bypass in a child with unsuspected Duchenne muscular dystrophy.

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Introduction: Of the 150 cases reported in the pediatric perioperative cardiac arrest registry, 10 were associated with hyperkalemia at the time of arrest. No data was available to confirm the presence of a myopathy. (1) In an earlier study of pediatric cardiac arrests (n =25) reported to the North American Malignant Hyperthermia Registry, unrecognized myopathy was present in 48% of patients (n=12) with hyperkalemia being present in 8 of the 12 cases (2). Malignant hyperthermia occurred only in 5 patients prior to the arrest. We report an uncommon cause of hyperkalemic cardiac arrest following cardiac surgery on cardiopulmonary bypass.

Case Report: A 4-year old 16.4 kg, otherwise healthy, hispanic male child presented for surgical closure of a large secundum atrial septal defect (ASD). There was no family history of congenital heart disease, early deaths, or anesthesia related problems. Pre-operative echocardiogram revealed a 14mm secundum ASD, with slight enlargement of the right heart. The child was premedicated with oral midazolam and anesthesia was induced with sevoflurane in oxygen and nitrous oxide. Neuromuscular blockade was achieved with pancuronium prior to nasotracheal intubation. Anesthesia was maintained with fentanyl (15mcg/kg) and isoflurane given both on and off bypass. Total bypass and cross clamp times were 29 and 14 minutes respectively. The heart was arrested with 25cc/kg of cardioplegia solution (7.5 mEq K+/L). During and following bypass, the patient was not hyperthermic, acidotic, or hyperkalemic (K+ ranged from 2.94 to 3.71 mEq/l). On arrival to the cardiac intensive care unit, his neuromuscular blockade was reversed and his trachea extubated. Twenty minutes later, the EKG showed broad complex bradycardia that rapidly progressed to ventricular tachycardia and fibrillation, and responded to defibrillation (20 J). The patient was re-intubated and the K+ found to be > 9 on the first arterial blood gas. The child's esophageal temperature was 37.7 ºC. Patient received calcium gluconate, sodium bicarbonate and was begun on an insulin and glucose infusion. To rule out rhabdomyolysis, creatine kinase (CK) was sent within an hour of the arrest showing a CK of 17,821 U/L. The CK peaked at 613,120 U/L 48 hours post-operatively. The patient was again extubated on the 1st post-operative day and improved with conservative management to be discharged to home on the sixth post-operative day. During further questioning of the family in Spanish, it was determined that the child did not walk until age 2 years. Genetic testing of a peripheral blood sample demonstrated a deletion in the dystrophin gene from at least exon 47 to exon 52 (exons 45 and 60 were present) that was consistent with a diagnosis of Duchenne muscular dystrophy (DMD).

Discussion: Adverse reactions to volatile anesthetic agents (3,4) and depolarizing muscle relaxants occur in DMD resulting in acute rhabdomyolysis and hyperkalemia, sometimes with clinical manifestations of malignant hyperthermia. Usually, features of a hypermetabolic state, rhabdomyolysis and hyperkalemia occur within a short period after exposure to volatile agents. In our patient, the manifestation was delayed by 3 hours and there were no features to suggest a hypermetabolic state.
Neonatal screening, by adding an inexpensive CK test to the specimen now obtained for inborn metabolic error screening could potentially diagnose all cases of DMD and other myopathies. (2) Avoidance of known triggers is essential for safe practice of anesthesia in patients with DMD. It has also been suggested that during peri-anesthetic arrests, the pediatric ALS protocol, be modified to detect and treat hyperkalemia. (2) A history of motor delay in children should be thoroughly investigated, and we suggest that serum creatine kinase levels be determined during the pre-operative evaluation, in this group of children.

References:

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