Malignant Hyperthermia in a two-month-old Infant During Cardiac Surgery: A Case Report
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Introduction: Malignant Hyperthermia (MH) is rare in infants younger than one year of age and the youngest confirmed by in vitro contracture testing (IVCT) was 6 months. MH is known for extremely high creatine phosphokinase (CK) levels, metabolic acidosis, muscle rigidity, and high temperature. However, MH has atypical presentations and if not treated aggressively, has a high mortality. Although high CK characterizes MH, it is a weak marker as patients have demonstrated mildly elevated or normal CK after MH.

Case Report: We present a 2 month old for repair of atrial and ventricular septal defects. She has a balanced translocation between chromosomes X and 2 at bands Xp21.2 (several important genes are in this region, specifically Duchenne Muscular Dystrophy) and 2q37.3 respectively. There was no family history of anesthetic problems. Anesthesia was induced with etomidate and vecuronium and maintained with fentanyl, midazolam, and sevoflurane. After uneventful bypass her temperature increased from 37 to 40.2 degrees C despite aggressive cooling. An arterial blood gas showed pH 7.26, PaCO2 54, base excess 2, and potassium increased from 3.6 to 5. Her PaCO2 increased and then was controlled by continuously increasing minute ventilation from 1 to 1.9 L/min. Two doses of dantrolene (1mg/kg) were given and the temperature decreased rapidly from 40.2 to 38.3. In the CTICU, her temperature increased again to 40. She was treated with dantrolene every 6 hours for 30 hours. CK values were 1152, 1308, and 1380 at 1, 2, and 12 hours after surgery. A metabolic acidosis was present with a base deficit of 5.3 at 4 hours after surgery. After 2 days her acidosis resolved and urine output remained clear.

Discussion: Our patient had a raw score of 35 on the Malignant Hyperthermia grading scale and a rank of 5 indicating that MH was very likely. MH is considered very likely in this case despite features being absent: muscle rigidity, myoglobinuria, and family history. The CK was high but only slightly more than expected for the surgery itself. Although there are risks with dantrolene, they are rarely life threatening and the benefits here outweigh the risks. A variety of physiologic changes can happen simultaneously after bypass that can confound the picture. The presence of central venous and arterial lines, inotropes, and frequent arterial blood gas analysis help treat changes more rapidly and effectively than if the episode occurred during non-cardiac surgery. Dantrolene was given because the temperature and CO2 responded to no measure and that it was low risk. Dantrolene rapidly decreased the temperature and CO2 which highlights its potential benefit in an otherwise unspecified refractory hyperthermia. A muscle biopsy could confirm the diagnosis and she could be the youngest. The presentation of MH can be variable. CK values may be mildly elevated, and dantrolene should be used early even when the diagnosis is unclear.

REFERENCES