Delayed Malignant Hyperthermia after congenital heart surgery
Michelle Sheth1, M.D.  Clair E. Nettles2, M.D.  Daniel J. Dibardino3, M.D. and Jorge D. Salazar3, M.D.
1. Department of Anesthesiology, 2. Department of Pediatrics, 3. Department of Surgery
Children’s Heart Center, The Blair E. Batson Children’s Hospital University of Mississippi Medical Center, Jackson, MS

Introduction

Malignant Hyperthermia is a rare life-threatening condition that is usually triggered by exposure to certain drugs used in general anesthesia specifically the volatile anesthetic agents and the neuromuscular blocking agent succinylcholine. In susceptible individuals, these drugs can induce a drastic and uncontrolled increase in skeletal muscle oxidative metabolism which overwhelms the body's capacity to supply oxygen, remove carbon dioxide and regulate body temperature eventually leading to a mixed metabolic and respiratory acidosis, circulator collapse and death if not treated quickly.

In the pediatric population, the occurrence rate is 1 in 15,000-2. Even rarer is delayed onset of symptoms with only a few documented cases in the literature 3. In this case report the onset is delayed which is extremely rare and early diagnosis may be missed with confounding factors such as bypass cooling. Early suspicion of malignant hyperthermia must be made when there is postoperative hyperthermia.

Case Presentation

A symptomatic infant female referred for surgical repair of a moderate sized perimembranous ventricular septal defect. She received an inhalation induction with a mixture of oxygen/nitrous with sevoflurane. Maintenance of anesthesia with fentanyl, vecuronium, and dexametomidine. Sevoflurane was used throughout the case. Through median sternotomy she was placed on cardiopulmonary bypass and underwent uneventful VSD closure and transferred to the pediatric cardiac ICU in stable condition.

The next morning, she was tachycardic with heart rate in 190s but was otherwise stable. At 10:00 AM, nearing 24 hours post-op, she had a fever to 106.1 that was recalctrant to antipyretics. ABG indicated profound mixed acidosis with pH of 7.15. Acidosis worsened despite fluid therapy and ice packs, and she was reintubated at 4.45 am. CK was ordered due to concern for a late onset malignant hyperthermia. Markedly elevated at >20,000 units/L, a Dantrolene bolus followed by infusion for 72 hours was started. She became afebrile soon after initiation of infusion with concomitant decrease in CK to a low of 61 units/L. All cultures were negative and genetics for RYR1 and 2 were obtained with results pending.

Discussion

The onset of malignant hyperthermia in our patient was delayed. The first symptom was tachycardia 18 hours post-op, followed by hyperthermia 24 hours post-op, and acidosis 30 hours post-op. During this time period, there was concern that the fever and acidosis were due to inflammation resulting from the body’s response to by-pass. This can be a confounding factor in post-cardiac surgery patients3. A diagnosis of malignant hyperthermia during bypass surgeries is difficult and can be delayed due to the cooling required for bypass4.

Due to the confounding factors, malignant hyperthermia should be considered in any child who presents with significantly elevated temperature and worsening acidosis post cardiac bypass procedure, regardless of time distant from procedure so that treatment may be started quickly.

References