[NM-224] Malignant Hyperthermia without Metabolic Acidosis

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Malignant hyperthermia (MH) is a rare, life-threatening disorder of skeletal muscle hypermetabolism in response to halogenated anesthetics and/or succinylcholine.
MH is linked to an inherited mutation that affects myocyte ryanodine receptors, resulting in rapid influx of calcium when exposed to triggering agents. This leads to intense muscle contraction and destruction, with subsequent chemical, thermal, and circulatory instability(1). Despite the expansive body of knowledge on MH, the diagnosis can be challenging. We present the case of a 13-year-old male with signs suggestive of, but not definitive for, MH.

A previously healthy 13-year-old, 77-kg, male presented for open repair of a right tibial tubercle fracture. He had no prior exposure to anesthesia, and family history was unremarkable.

After uneventful propofol induction and intubation without neuromuscular blockade, anesthesia was maintained with sevoflurane. Twenty minutes after induction, he became progressively tachycardic and hypertensive despite morphine and increased sevoflurane. The end-tidal CO2 rapidly climbed from 40mmHg to 70mmHg despite upward adjustments in ventilation, and the temperature rose to 39 degrees Celsius. No muscle rigidity was present.

Concern for MH was declared. Sevoflurane was discontinued and a propofol infusion initiated. Manual hyperventilation with 100% FiO2 was attempted, but the patient had spontaneous respiratory drive with a minute ventilation of 19 L/min on pressure support. A bladder catheter demonstrated clear urine, and he was aggressively fluid resuscitated. Active cooling was initiated. Venous blood gas showed respiratory acidosis without metabolic acidosis.

With these interventions the end-tidal CO2, HR, BP, and core temperature began to normalize. The MHAUS hotline was consulted and advised holding off on dantrolene administration given the lack rigidity, lack of metabolic acidosis/base excess, and evidence of symptom resolution. The procedure was rapidly completed, and the patient emerged and transferred to the PACU. He developed postoperative myoglobinuria with a rise in CK from 4,565 to 14,584 U/L on POD #2. Serum myoglobin was 8,095 ng/mL. Additional questioning revealed that the patient and a 3-year old biological sister had always been unusually muscular since birth. He frequently suffered from post-exercise muscle cramping. Therefore, a putative diagnosis of RYR1 myopathy was entertained, and the patient was counseled on avoidance of anesthetic triggering agents until further diagnostic work-up occurred.

1. Clinical Presentation, Treatment, & Complications of MH in N. America from 1987 to 2006. Int Anes Res Soc 2010; 498-507

Venous Gas Sample	Initial Values	1 Hour post	48Hour post
pH	7.21	7.39	-
PvCO2 (mmHg)	70	47	-
$HCO3^-$ (mEq/L)	28	28	-
BE (mEq/L)	-1	2.5	-
K^+ (mEq/L)	4.9	5.0	-
Creatine Kinase (U/L)	4,565	-	14,584
Serum Myoglobin (ng/ml)	8,095	-	-