A General Anesthetic with Neuromonitoring in a Patient with MERRF
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Introduction
MERRF (myoclonic epilepsy with ragged red fibers) is a rare, maternally inherited mitochondrial disease affecting enzyme complexes I and IV of the respiratory chain).

Features include:
- Myoclonic epilepsy
- Ataxia
- Hearing loss
- Elevated serum lactate

Organ systems that rely on aerobic metabolism are most affected, specifically:
- The central nervous system
- The cardiovascular system
- Skeletal muscle

Mitochondrial diseases have widespread systemic effects and can be exacerbated during periods of stress. For this reason, the perioperative care of children with MERRF presents unique challenges.

Case
An 11 year-old girl presents for posterior spinal fusion. She was diagnosed with MERRF and Leigh syndrome at age 6 after presenting with hearing loss and ataxia, and was gastrostomy tube dependent due to loss of tongue control. Preoperatively, she was evaluated by Endocrinology, Neurology, Cardiology, and Pulmonology specialists with pertinent findings including a serum lactate of 4.6 mM/L and medication-induced hyponatremia and hypochloremia. In the past, the patient’s symptoms had worsened while NPO, thus she was admitted preoperatively for initiation of parenteral nutrition.

Anesthesia was induced with midazolam, fentanyl and ketamine, and maintained with remifentanil, midazolam, ketamine, and dexmedetomidine infusions. Her parenteral nutrition was continued intraoperatively. Baseline labs showed pH 7.36, lactate 2.6, Na 125, Cl 92, and glucose 179. Serial ABGs, lactate, electrolytes, and glucose levels were followed. At the end of surgery her lactate was stable at 2.1, pH was 7.35, sodium had normalized to 132, and chloride was 111. Blood glucose remained in goal range. She was brought to the PICU intubated for post-operative care and was extubated on post-operative day 2.

Discussion
MERRF influences several aspects of anesthetic care. Preoperative evaluation should assess for known sequelae of mitochondrial disease including cardiomyopathy and arrhythmias. Anesthetic goals in the management of patients with MERRF include:
- Normothermia
- Normotension
- Adequate oxygenation
- Normoglycemia
- Avoidance of factors known to precipitate lactic acidosis

We used several strategies to achieve these goals including:
- Intraoperative glucose supplementation (continuing TPN)
- Close glucose monitoring
- Dexmedetomidine infusion instead of propofol
- Normal saline instead of lactated ringers

There is controversy regarding increased risk of malignant hyperthermia in mitochondrial disease. In this case, the need for intraoperative neuromonitoring necessitated delivery of a general anesthetic without volatile agents. Ketamine and remifentanil were used together for intraoperative analgesia.

References

Figure 1. Mitochondrial electron-transport chain, taken from Figure 1 Pediatr Anesth 2013; 23: 786.

Figure 2. “Ragged Red Muscle Fibers” with Gomori Trichrome Stain
http://neuromuscular.wustl.edu/pathol/mitochondrial.htm#rrf

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