[NM-206] Anesthetic Management and Concerns in a Child with Pelizaeus-Merzbacher Disease

[1]Fabian J, [2]Pryjdun V, [2]Christofaro R, [2]Barst S
[1]New York Medical College , Valhalla , New york, USA; [2]Westchester Medical Center / New York Medical College , Valhalla , New york, USA

Summary: We present the case of a child with Pelizaeus-Merzbacher disease (PMD), scheduled for bilateral femoral osteotomies. We briefly discuss the concerns involved in the anesthetic management of a patient with this rare disorder.

Case Report: The 8 year-old patient was diagnosed antenatally by amniocentesis (positive family history of PMD in a maternal uncle). PMD is one of the leukodystrophies characterized by progressive degeneration of myelin that is X-linked and the result of a mutation in the proteolipid protein gene (PLP1). The clinical presentation and severity of the disease vary widely, but patients typically present in infancy with cognitive delay, spasticity, weakness, nystagmus, and seizures. PMD often follows a progressive, degenerative course, which may include periods of relative stability.

The patient arrived in the OR holding area the morning of surgery. NPO status was confirmed. Past medical history was notable for global developmental delay (motor, cognitive and language milestones) and mild gastroesophageal reflux. Past surgical history included correction of bilateral clubbed-foot deformities.

Standard ASA monitors were applied and 22g IV access established in each upper extremity. The patient was pre-oxygenated and induced with propofol, fentanyl, and sevoflurane. The airway was secured with a MAC #2 and a 5.5 cuffed endotracheal tube; placement was confirmed with ETCO2. On orogastric tube was placed, the patient received glycopyrrolate, and a forced hot-air heating blanket was used to prevent hypothermia. Intraoperative fluids included Plasma-Lyte A 1800 mL and 1 unit of packed red blood cells. Urine output was 100 mL. The patient was maintained on sevoflurane (2-3%) and pressure-mode ventilation: PAP 15 cm H2O, PEEP 4 cm H2O, and VTE 250 mL at RR of 12. Surgery was uneventful and the patient was extubated after demonstration of intact airway reflexes. He was transported to the PACU with facemask O2 and pulse O2 monitoring. The patient remained in the PACU for postoperative monitoring as he was weaned to room air.

Discussion: Possible concerns during anesthetic management of a patient with PMD leukodystrophy include an increased risk of aspiration, increased oral secretions, complications of airway management related to CNS and muscle control, increased sensitivity to respiratory depression, increased potential for hypothermia, and seizures.
We elected to avoid the use of nondepolarizing muscle relaxants due to possible prolongation of neuromuscular blockade in patients with demyelinating diseases. We elected to avoid succinylcholine because of a risk of potential hyperkalemia, although we could not find literature to support this concern. Meticulous ventilator care with the early initiation of PEEP, appropriate tidal volumes and peak airway pressures to prevent respiratory decompensation should be used. In the care of any child with multiple complex problems attention to detail and appropriate back-up is essential for a safe outcome.

References
Baum, VC, O'Flaherty JE. Anesthesia for Genetic, Metabolic, and Dysmorphic Syndromes of Childhood. Lippincott Williams & Wilkins: 2007; 205-6
