Challenges in the Anesthetic Management of a Pediatric Patient with Pelizaeus-Merzbacher Syndrome

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Introduction
Pelizaeus-Merzbacher syndrome is a rare X-linked recessive genetic disease characterized by progressive unvarying demyelination of the central nervous system, which manifests as developmental delay, muscle tone abnormalities (spastic or hypotonic), seizures, nystagmus, dysphagia, and respiratory collapse. Due to the infrequency of this disorder in the general population, there is scarce literature describing the concerns for anesthetic management, particularly of a pediatric patient. This case report reviews the challenges and considerations in the perioperative management of a three year-old child with Pelizaeus-Merzbacher syndrome undergoing general anesthesia for outpatient dental restoration.

Case Report
Our patient is a three year-old male with Pelizaeus-Merzbacher syndrome weighing 14 kg, who at baseline has generalized hypotonia and requires the use of a wheelchair, however can use braces to ambulate with assistance. He has bilateral nystagmus on neurologic exam, yet there was no history of seizure activity, dysphagia, or respiratory difficulties. The patient underwent an inhalational induction with nitrous oxide and sevoflurane, which he tolerated with parental presence. Intravenous access was established, and endotracheal intubation was achieved with an uncuffed nasal Rae. He received multimodal analgesia with fentanyl and acetaminophen, as well as dexamethasone, ondansetron, and dexmedetomidine. Prior to extubation, the patient had adequate tidal volumes breathing spontaneously and baseline strength was observed in the head and all extremities. He was transported to the PACU in stable condition and discharged home later that day.

Discussion
Given the neurologic manifestations of Pelizaeus-Merzbacher, there are numerous implications to modification of anesthetic management. Premedication with an oral benzodiazepine was avoided due to the concern for increased sedation in a hypotonic patient, placing him at risk for aspiration. Patients with this disorder who have a significant seizure history may however find it beneficial to employ the use of benzodiazepines. Succinylcholine was avoided during induction given this patient’s increased risk of a hyperkalemic response, and nondepolarizing muscle relaxation was also avoided to prevent any exaggerated hypotonia at completion of the case. Weakness of oropharyngeal musculature often place these patients at a high risk of aspiration, both in the operating room and at home. With a detailed history and physical exam, it was determined that this patient did not have a significant history of dysphagia or increased salivation. Multimodal analgesia with limited short-acting opioid as well as dexmedetomidine administration was used to prevent any postoperative respiratory depression. Ventilation goals included normocapnia, with early initiation of peep, adequate tidal volumes and low peak airway pressures to prevent postoperative respiratory complications. Hypothermia was avoided with forced-air patient warming to prevent seizure activity and further weakness. The patient remained hemodynamically stable with crystalloid to achieve normovolemia and minimal blood loss.

Overall, perioperative management of patients with Pelizaeus-Merzbacher requires a comprehensive anesthetic plan. By anticipating any complications, patients with this disorder can successfully undergo surgical procedures with general anesthesia.