

# [NM-141] Anesthesia Management in a Patient with Leigh's Syndrome and a Family History of MH Presenting for Dental Rehabilitation

<sup>1</sup>Edala T, <sup>2</sup>M I, <sup>2</sup>Edwin A, <sup>2</sup>Siddiqui S, <sup>2</sup>Jesus A, <sup>1</sup>Edala T

<sup>1</sup>Uams , Little Rock , Arkansas, USA; <sup>2</sup>Arkansas Children's Hospital , Little Rock , Arkansas, USA

---

Leigh's Syndrome is a rare disorder caused by mutations in mitochondrial DNA or by deficiencies of pyruvate dehydrogenase enzyme (1,2). Providing these patients with the most suitable anesthetics remains problematic.

## Case Report

A 19-year-old male with Leigh's Syndrome was scheduled for dental rehabilitation. Past surgical history included a gastrostomy and Nissen fundoplication under general anesthesia at an outside facility. Details about the anesthetics were not available. Family history was positive for a first cousin with hypotonia of unknown diagnosis and masseter spasm on exposure to anesthesia and death unrelated to the episode. Patient was the only sibling to a single parent. An echocardiogram reported cardiac dextroposition due to severe pectus carinatum deformity & normal biventricular functions.

As patient's incomplete history was suspicious for malignant hyperthermia susceptibility, total intravenous anesthesia was planned. Preoperatively, midazolam was administered subcutaneously. Anesthesia was started with 70% N<sub>2</sub>O in O<sub>2</sub> with vapor free anesthesia machine. Following placement of a peripheral IV, a bolus of dexmedetomidine (1 µg/kg) over 10 minutes and fentanyl was given before nasal endotracheal intubation. Anesthesia was maintained with oxygen & nitrous oxide and continuous infusions of dexmedetomidine (0.4 - 1.4 µg/kg/hour) and remifentanyl (0.8 - 1.2 µg/kg/min). Normal saline was used as maintenance fluid. The postoperative course was uncomplicated and he was discharged home with advice to take ibuprofen.

## Discussion

The rarity of patients with Leigh's Syndrome results in a paucity of information regarding anesthesia management. Although all anesthetics have been shown to moderately suppress the mitochondrial respiratory chain, both propofol and sevoflurane have been used without problems (3,4). However, most experts agree avoiding propofol in patients with a known diagnosis of mitochondrial disease (4,5,6). Although malignant hyperthermia has not been reported with Leigh's Syndrome (5), it may be best to avoid triggering agents as other myopathic conditions with susceptibility for malignant hyperthermia may be misdiagnosed as Leigh's Syndrome. In this patient who had a relative suspicious for malignant hyperthermia susceptibility, we chose to use dexmedetomidine with remifentanyl for the induction and maintenance of anesthesia. For reasons mentioned earlier, these authors decided to use TIVA but without using propofol. Both of these drugs have rapid recovery profile, limited postoperative CNS and respiratory depression that made them well suited for this case. It is also recommended avoiding circumstances that place a metabolic burden on these patients like prolonged fasting and pain, nausea and vomiting, hypothermia, and prolonged tourniquets. Saline instead of lactated ringers is advocated.

## References:

1. Leigh D. J Neurol Neurosurg Psychiatry 1951; 14:216–21.
  2. Morgan PG, et. al. Anesthesiol 2002; 96:1268–70.
  3. Finsterer J, Segall L. Drug Chem Toxicol 2010;33:138-51
  4. Niezgoda J and Morgan P. Pediatr Anesth 23 (2013) 785–793
  5. Kinder Ross A. Pediatr Anesth 2007; 17: 1–6.
  6. Driessen JJ. Curr Opin Anaesthesiol. 2008;21:350–5.
-