Anesthetic management of a pediatric patient with Leigh syndrome during Magnetic Resonance Imaging

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Introduction

Leigh syndrome, also known as subacute necrotizing encephalomyelopathy, is a rare progressive neurodegenerative disorder affecting only 1:40,000 and associated with mitochondrial DNA mutations in about 20% of those diagnosed with the disorder. Research has led to limited case reports and few reviews discussing the anesthetic management of Leigh syndrome. The purpose of this report is to yield a comprehensive review of pediatric anesthetic management of a patient with suspected Leigh syndrome, and particularly management during MRI since this imaging is used in diagnosis of the syndrome.

Case Description

A 4 year-old male with significant medical history including encephalopathy, hypotonia, and developmental delay presented to the MRI suite for diagnostic evaluation of suspected Leigh syndrome.

The patient had a medically complex history including hypoxic injury shortly after birth followed by possible seizures. Past medical history includes encephalopathy, progressive worsening developmental delay, and G-tube dependence with increasing intolerance to feeds. The patient had no previous anesthetic record at the hospital but no history of problems with anesthesia per parents, including safe use of volatile anesthetics. Careful assessment of respiratory and cardiac function was conducted including a full history and exam. Information was limited since the patient had not received care at this hospital prior to the MRI.

He was cooperative with mask induction after the G-tube was suctioned and vented. IV placement was difficult but obtained without any changes in hemodynamics during that time. The patient was then intubated and transported from the induction area to the MRI. In the MRI, he was maintained on low dose sevoflurane at 0.5 MAC and a dextrose infusion of intravenous fluids. He was extubated awake and eventually discharged after ensuring normoglycemia without any anesthetic complications.

Discussion

Leigh syndrome is categorized as part of the mitochondrial myopathies, even through only 20% of these patients have associated mitochondrial DNA mutations. Neurologic features include hypotonia, spasticity, movement disorders, cerebellar ataxia, and peripheral neuropathy. Patients may have hypertrophic cardiomyopathy and can die to respiratory or cardiac failure. While many different anesthetic techniques have been successful in this population, serious complications, such as episodes of lactic acidosis and respiratory failure, can occur. This results in a general opinion that these patients are at increased risk from anesthetic exposure.

It is important to obtain a comprehensive history and exam in order to understand the extent of the disease and whether the patient has any respiratory, cardiac, or other concerns that may increase morbidity. Previous literature suggests avoidance of volatile agents, propofol, lactate containing fluids, and certain neuromuscular blocking agents. However, cases have shown mixed results when using these agents usually resulting in either no complications or typically respiratory complications. Maintaining hemodynamic stability, normocapnia, and normothermia are imperative. Dexmedetomidine and remifentanil infusions have been safely used for maintenance of anesthesia in these patients.

Often it is not possible to know the underlying genetic defect causing mitochondrial myopathies such as Leigh disease at the time of the encounter. Thus, the anesthesiologist should make the best interpretation of the available data in order to create an anesthetic regime that will lower the risk of complications of an already compromised patient. Further discussion should be considered to analyze the utility of small doses of volatile agents and propofol infusions for minor non-stimulating procedures such as a skin biopsies or imaging studies.

Anesthetic considerations:

1) Minimize preoperative fasting to avoid hypovolemia, hypoglycemia, and increased use of fatty acids
2) Careful preoperative cardiac and respiratory evaluation
3) Effective perioperative antiepileptic therapy and normal electrolytes
4) Cautious use of muscle relaxants in patients with a preexisting myopathy or decreased respiratory drive
5) Maintain normothermia
6) Avoid hypocapnia and hypoxia
7) Slow titration of volatile and parenteral anesthetics to minimize hemodynamic changes
8) Use measures to decrease PONV
9) Consider malignant hyperthermia precautions

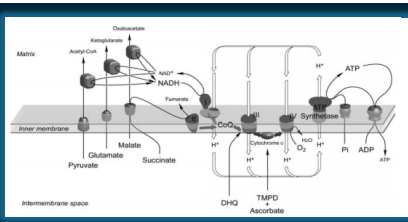

Figure 1: Defects in the mitochondrial respiratory chain can lead to changes in cellular metabolism in mammals.

References

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