

Congenital Hypotonic Syndrome with Persistently Elevated CPK: Anesthetic Concerns

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Introduction: Anesthetic management of hypotonic infants and children are fraught with the following questions: (1). Is this child somehow at greater risk for Malignant Hyperthermia?
(2). Is this child at greater risk for rhabdomyolysis if volatile anesthetic agents are used?
(3) What is the evidence for either of the above?
(4) Is Total Intravenous Anesthesia the best anesthetic choice for this child?

Case Presentation: An 18-month-old, 11 kg female was scheduled for muscle biopsy for congenital hypotonia syndrome. The child had global developmental delay. She was not able to stand up, but she could sit unsupported. MRI of the brain showed abnormal white matter. There was no family history of muscle disease. She was normal for amino acid disorders as well as for other metabolic disorders. Genetic work up showed no chromosomal abnormalities. A working diagnosis of Alpha 2 Laminin deficiency (Merosin) was being entertained by the child's Neurologist. Up until the presentation of the case there was no pathological findings suggestive of merosin deficiency, except the hypotonic morphology. Child's CPK at presentation was 800U/L. During a literature search we found a single case report of an infant with merosin deficiency who had been given a 'non-triggering anesthetic, and went on to develop what was described as MH during his anesthetic course(1). Following the lead of this somewhat confusing case report, we decided to provide a non-triggering anesthetic for our patient, with bolus doses of midazolam, fentanyl, and propofol infusion along with nitrous oxide. The anesthetic time lasted three times as long as the surgical time. The anesthesia and recovery was uneventful. Shortly after this anesthetic, further work up of this child revealed normal staining for Laminin alpha2, placing the child in the category of 'Congenital Hypotonia of Unknown Etiology'.

Discussion: It appears as though the practice of using total intravenous anesthesia for infants and children with congenital hypotonia of known or unknown origin has become the 'default' position(2). More detailed literature search for anesthetic management of these children do not reveal the rationale behind this approach nor does it appear to have 'evidence based' approach. We would also like to point out that each child with hypotonia/myopathy deserves individual consideration for their anesthetic requirement and not be branded into the 'Total Intravenous Anesthesia' group, especially considering that this group of patients may be at greater risk for propofol infusion syndrome(3,4). Not all hypotonic patients are the same and given each patient's unique diagnosis and underlying progression of the disease a suitable anesthetic should be considered.

Refs: (1). Suspected malignant hyperthermia in a child with Laminin alpha2 (Merosin) deficiency in the absence of a triggering agent. *Paediatr Anaesth* 2006; 16:462-5
(2) General anesthesia for a patient with centronuclear (myotubular) myopathy. *Anesthesiology* 1998; 89:1018-20.
(3) Impaired fatty acid oxidation in propofol infusion syndrome. *Lancet* 2001; 357:606-7.
(4) Acquired carnitine deficiency: a clinical model for propofol infusion syndrome? *Anesthesiology* 2005; 103:909.