**Risk of Malignant Hyperthermia in Children with Duchenne Muscular Dystrophy**

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**Introduction:** Duchenne muscular dystrophy (DMD), an X-linked recessive disorder has been associated with an increased risk of malignant hyperthermia (MH) in several anesthesiology textbooks and journal articles (1, 2, 3). DMD is found almost exclusively in males, occurs at a rate of approximately 1-3 cases per 10,000 live male births, and is ultimately fatal, usually by the end of the second decade of life. Diagnosis is typically made between the ages of 3-5 years and rarely after age 10 years. Although several reports have linked DMD with malignant hyperthermia, no large study has confirmed the relationship and it remains unclear whether increased risk actually exists. Anesthetic management of children with a known or suspected risk for the development of MH is complex, requiring preparation of the anesthesia machine and awake placement of intravenous access in order to avoid the use of volatile anesthetic agents. This population-based study examines the anesthetic management of children known to have DMD in an effort to determine whether those exposed to volatile anesthetic agents developed signs or symptoms consistent with malignant hyperthermia.

**Methods:** All Olmsted County, Minnesota residents with a medical diagnosis code of myopathy, Duchenne muscular dystrophy or any other progressive muscular dystrophy from 1957-1993 were identified. Charts of those with DMD who underwent surgery and anesthesia were retrospectively reviewed. The type of anesthesia, use of volatile anesthetics, use of depolarizing muscle relaxants, and adverse events including fever, cardiac arrest, masseter muscle spasm, rigidity, evidence of rhabdomyolysis, and death were identified.

**Results:** During the 37-year period, Duchenne muscular dystrophy was diagnosed in 23 patients who underwent 47 surgeries. Interestingly, one girl with DMD was identified who also carried the diagnosis of Turner Syndrome. The mean age at diagnosis was 5 years 6 months. All children were diagnosed by 9 years of age. Average duration of anesthesia was 147 minutes. Forty-four of 47 patients had general anesthesia, all with volatile anesthetic agents. The volatile agents were halothane in 21 patients, isoflurane in 19 patients, and enflurane in 4 patients. Prior to a diagnosis of DMD, 2 of 47 patients received succinylcholine. No adverse events were identified.

**Discussion:** Duchenne muscular dystrophy is a relatively rare disease yet has important anesthetic implications including malignant hyperthermia and succinylcholine-induced hyperkalemic cardiac arrest. In this retrospective study of 47 surgeries in 23 patients with DMD, in which 93% of anesthetics included volatile agents and 4% included succinylcholine, no adverse events were encountered. No child was found to have fever, evidence of masseter spasm or rhabdomyolysis, or other signs of malignant hyperthermia. Neither child given succinylcholine developed electrocardiographic evidence of hyperkalemia.

In this small study, no patients with DMD developed evidence of malignant hyperthermia despite having received halogenated volatile anesthetics. In the two patients who received succinylcholine prior to DMD diagnosis, no evidence of hyperkalemia or malignant hyperthermia was identified. However, we do not advocate the use of succinylcholine in these patients due to the well-established increased risk of hyperkalemic cardiac arrest.

**References:**

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