**CLINICAL COMMENTARY**

# Duchenne Muscular Dystrophy and Anesthesia

**Fei Zheng-Ward, M.D.**

Duchenne muscular dystrophy (DMD) is an X-linked recessive disorder that affects approximately 1 in 3,500 males worldwide.[1,12] It is due to an abnormal dystrophin gene on the X chromosome that results in the lack of dystrophin production. Dystrophin normally provides mechanical reinforcement to the sarcolemma of the muscle tissue and stabilizes the glycoprotein complex. In its absence, the glycoprotein complex is digested by proteases and may initiate the degeneration of muscle fibers, resulting in muscle weakness. Clinical onset of muscle weakness typically manifests between 2-5 years of age.[2]

The condition is characterized by waddling gait, lumbar lordosis, calf enlargement, weakness, falls, and atrophy of the various muscle groups.[3,11] The pattern of muscle weakness usually involves the proximal limb muscles earlier than distal and lower extremities before upper. Other organ systems that are involved include neurologic, cardiac, respiratory, and skeletal, which can manifest as mental retardation, cardiomyopathy, respiratory insufficiency, and scoliosis to name a few.

With advances in medicine, patients with Duchenne muscular dystrophy are living longer and are presenting to the hospital for multitude of surgical procedures. As a result, it is important to understand the anesthetic management of these patients in the perioperative periods.

The anesthetic management depends on the organ systems affected by the disease. Due to the risk of hyperkalemic cardiac arrest and rhabdomyolysis, succinylcholine is contraindicated. The incidence of malignant hyperthermia (MH) is also increased in these patients and Dantrolene should be readily available.[3,7] However, the use of volatile agents is controversial and should be determined on a case-by-case basis. Brief use of volatile agents for securing a difficult airway is reasonable, but most providers would switch to total intravenous anesthesia (TIVA) for the duration of the surgical case. For patients who are exposed to volatile agents, it is reasonable to monitor them for signs of rhabdomyolysis by checking serial serum potassium and creatinine kinase (CK) levels.[4] Nonetheless, despite monitoring complications may still occur with use of volatile ***and*** nonvolatile agents.[5]

***Preop Assessment***

DMD not only affects the skeletal muscles, but it can have an adverse effect on the myocardium due to dystrophin deficiency that can result in progessive cardiomyopathy. Thus, preanesthesia evaluation should include cardiac evaluation and/or consultation.[6] Additionally, as respiratory system is often involved, some providers opt to obtain baseline respiratory parameters. The disease process can pose significant risks on these organ systems in DMD patients and increase their morbidity and mortality while they undergo major surgeries.

***Intraop Management***

Consider flushing the anesthesia machine prior to use on DMD patients and use a clean circuit, filters, and brand new CO2 absorbent canister. Avoid succinylcholine due to the risk of rhabdomyolysis and hyperkalemic cardiac arrest. Some providers suggest not using muscle relaxants and reversal to reduce postoperative respiratory failure; however, this is not always possible for some surgical procedures. There are case reports of successful use of sugammadex in DMD patients without residual curarization.[8] Additionally, respiratory parameters should be checked and compared to preoperative values, if available, prior to extubation. If the patient needed continued intubation after surgery for an extended period of time, one may consider not using a reversal agent.

***Postop Management***

If rhabdomyolysis is suspected, then obtain serial plasma potassium, CK and myoglobin and urine myoglobin. Close monitoring may be indicated for DMD patients depending on the surgery and their medical condition.

Lastly, general and neuraxial anesthesia have been associated with high risks of complications that include, but are not limited to, rhabdomyolysis, hemodynamic instability, malignant hyperthermia, respiratory depression, and postoperative mechanical ventilation.[9] Peripheral nerve blocks are usually safe and reasonable for DMD patients undergoing orthopedic surgery, however, MH-like reaction is still possible so close monitoring may be indicated.[10,11]

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