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Anaesthetic Consideration in Patient with Duchenne Muscular Dystrophy - A Case Report

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Abstract: ***Duchenne Muscular Dystrophy (DMD) is caused by an X - linked recessive mutation in the dystrophin gene, leading to a lack of dystrophin, a crucial protein in muscle cell membranes. Patients with DMD suffer from significant muscle weakness and associated medical conditions such as cardiomyopathy, respiratory failure, and scoliosis. Effective anesthetic management requires thorough preoperative evaluation, advanced airway equipment, aspiration precautions, and careful patient positioning. Though not specifically at risk for malignant hyperthermia, DMD patients are susceptible to anesthesia - induced hyperkalemia, rhabdomyolysis, and cardiac arrest. This case study details the successful anesthetic management of a 12 - year - old male with DMD undergoing foreign body removal from the left ear under general anesthesia.***

Keywords: Duchenne muscular dystrophy ¸ difficult airway, muscle relaxant, anesthesia

1. Introduction

Muscular dystrophies are a heterogeneous group of diseases that share a common etiology due to direct injury of the muscle fiber with a progressive and systemic compromise. Each type of muscular dystrophy is different in terms of its clinical presentation, genetic origin, and anesthetic risks which are mainly cardiovascular complications, including malignant arrhythmias, acute rhabdomyolysis triggered by drugs used in anesthesia, and perioperative respiratory failure. These patients have difficult airway anatomy in the form of macroglossia and limited mobility of the mandible, cervical spine.

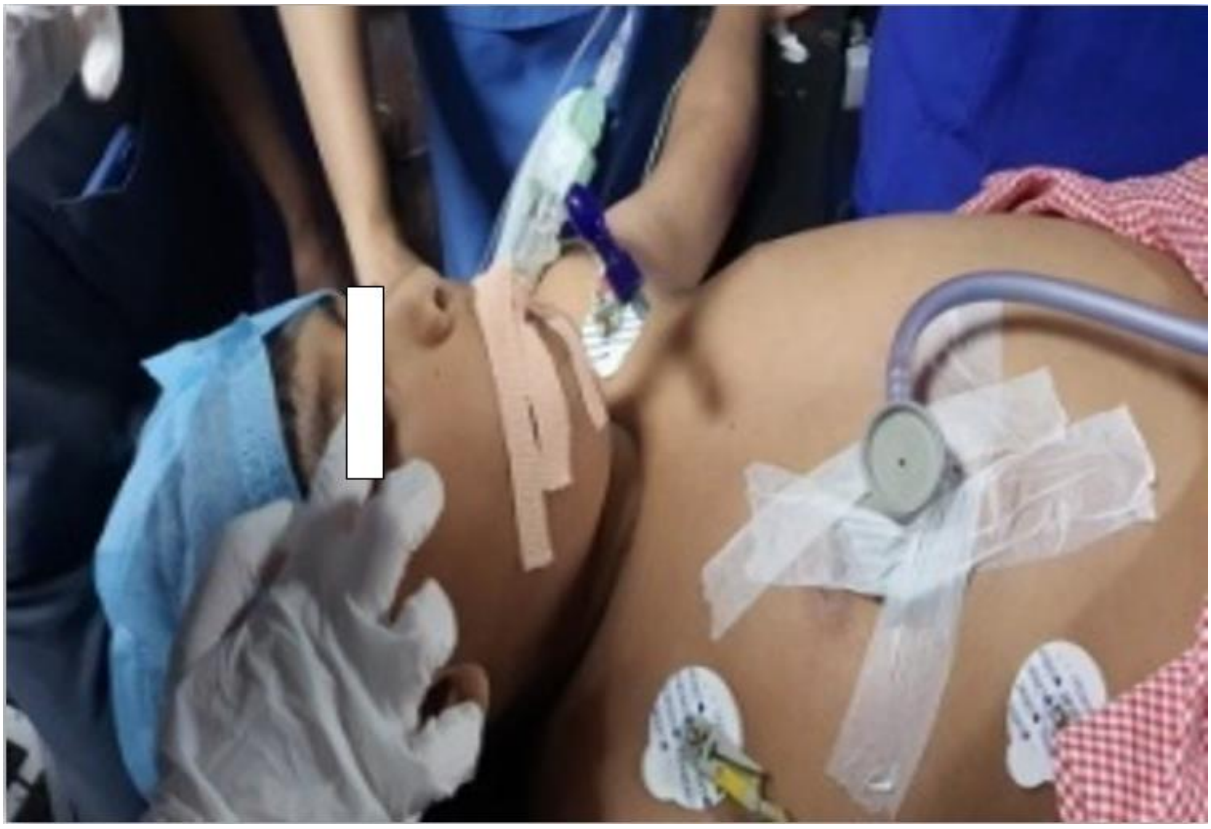
The purpose of this article is to detail the anesthetic management of a patient with DMD undergoing a surgical procedure, and emphasizes the importance of specialized anesthetic techniques and thorough preoperative assessments in managing patients with DMD to prevent potentially fatal complications.

2. Case Report

A 12yr old male child, weighing 35kg diagnosed with foreign body in left ear was scheduled for its removal. The child had difficulty in walking since the age of 3years with increasing bilateral lower limb weakness. Lab values showed **Serum CPK - NAC: 9645U/L**, and the pathogenic variant causative of the suspected phenotype was identified as a **hemizygous deletion of 3 Exons 48 - 50**. The child was diagnosed with duchenne muscular dystrophy at 8yrs of age and was advised treatment with Tablet Prednisolone 5mg once daily and Physiotherapy. Airway examination revealed a **larger tongue with Mallampatti grade III**. CNS examination showed decreased tone and power of 2/5 in all four limbs, calf muscle hypertrophy, and absence of knee reflex, and scoliosis while walking. Wheel chair bound since one year.

Patient was taken for the procedure under GA with high risk consent.
Standard ASA monitors were attached. The patient was preoxygenated with 100% 02 for 3minutes. Premedication included Inj Glycopyrrolate (0.01mg/kg) 0.2mg IV, Inj Midazolam (0.01mg/kg) 0.25 mg IV, Inj Fentanyl (2mcg/kg) 50mcg IV and Inj Hydrocort (5 mg/kg) 100mg IV. Induction was performed with Inj Propofol (2mg/kg) 50mg IV and followed by relaxation with Inj Atracurium (0.5mg/kg) 20mg IV. Airway was secured with a size 2.5 LMA and was maintained with 02 and N20. A bolus dose of Inj Ketamine (2mg/kg) 50mg IV was given.

A **precordial stethoscope** was placed throughout the surgery



Intraoperative was uneventful. The Patient was extubated after meeting extubation criteria and was shifted to the recovery room

3. Discussion

Children with neuromuscular disease commonly present for anesthesia as part of diagnostic process such as MRI or Muscle biopsy, for surgery relating to underlying disorder (gastrotomy, corrective orthopedic procedure etc) or for incidental surgeries. Incidence – 1: 3500 live male birth. Due to the absence or abnormal dystrophin there is a chronic muscle fiber necrosis, degeneration, and regeneration. Degeneration occurs in cardiac muscle, smooth muscle and skeletal muscle.

Infants at birth may appear normal at birth, weakness begins in childhood before age of 8 and is rapidly progressive. The muscle around the pelvis and thighs are affected first, presenting with difficulty in managing stairs and standing from sitting. By adolescence patient are usually wheel chair bound like the patient in this case report and they succumb to **cardiac or pulmonary manifestation** at their early 20s to 30s.

Anesthesia technique that minimizes cardiac and respiratory depression should be used utilizing drugs

that are short acting and rapidly metabolized. Most children with neuromuscular disease will have increased **sensitivity to depolarizing neuromuscular agents.**

Succinylcholine and inhalational agents should be avoided as they can cause life - threatening increases in serum potassium in response to Succinylcholine.

People with Duchenne are at risk of developing **rhabdomyolysis** (the breakdown of skeletal muscle tissue that may cause the release of myoglobin that can damage the kidneys) and **hyperkalemia** (the release of too much potassium into the bloodstream), which can result in life - threatening heart rhythms. There are known cases of serious (and sometimes fatal) muscle breakdown (rhabdomyolysis) in Duchenne patients when exposed to inhalation anesthetic gases even when Succinylcholine was avoided. **Malignant hyperthermia** can also be precipitated.

Motor and sensory neuromonitoring is the standard of care for these procedures even though the quality of baseline signals may be difficult to obtain due to muscle weakness.

A multidisciplinary approach is recommended for any surgery. The **safest anesthetic technique is an IV induction followed by maintenance with TIVA.**

4. Conclusion

Careful preoperative assessment and avoidance of depolarizing muscle relaxants and inhalational agents are crucial for successful anesthesia outcomes in patients with Duchenne Muscular Dystrophy. Maintaining normothermia and using short - acting, rapidly metabolized drugs can help prevent severe complications such as hyperkalemia and rhabdomyolysis.

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