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Anaesthetic Management of a Case of Duchenne's Muscular Dystrophy

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Abstract: ***A 9yr old male child, with Duchenne's muscular dystrophy was scheduled for tonsillectomy, adenoidectomy with bilateral grommet insertion. The anaesthetic management of patient with muscular dystrophies is complicated and challenging. A thorough preoperative evaluation and multidisciplinary approach is essential for perioperative management of such cases. Here, we describe the successful anaesthetic management of a case of Duchenne muscular dystrophy who underwent tonsillectomy, adenoidectomy & bilateral grommet insertion procedure under general anaesthesia.***

Keywords: Duchenne's muscular dystrophy, difficult airway. anaesthesia management

1. Introduction

Duchenne's muscular dystrophy is an X linked recessive disorder caused by a mutation in the dystrophin gene located on chromosome Xp21. Duchenne's muscular dystrophy affects males almost exclusively. Typical presentation is proximal muscle weakness in early stage of the disease. Patients develop respiratory distress and cardiomyopathy in the advanced stage of the disease.

Progressive weakness and contractures eventually result in kyphoscoliosis. The patients have associated difficult airway anatomy in the form of macroglossia, limited mobility of mandible and cervical spine. These patients are at increased risk of developing extreme hyperthermia, rhabdomyolysis & hyperkalemic cardiac arrest, when exposed to halogenated inhalational anaesthetics & depolarizing muscle relaxants.

2. Case Report

9yr old male child, weighing 24kg with Duchenne's muscular dystrophy was scheduled for adenoid & tonsillectomy, with bilateral grommet insertion. The child had progressively increasing bilateral lower limb weakness & was diagnosed to have Duchenne muscular dystrophy at the age of 5yrs. Child was on tablet prednisolone taking regularly & undergone stem cell therapy in the past 2 years.

Cardiovascular & respiratory systems were normal, Spo2 95% on air. The child has a history of mouth breathing & nasal obstruction due to adenoid. History of recurrent upper respiratory infection. Frequent falls, difficulty rising from a lying or sitting up position, trouble running and jumping, walking on the toes, large calf muscles, muscle pain and stiffness. Airway examination revealed a large tongue with Mallampatti grade 3, 2D Echo was normal with EF 65%. ECG was normal. MRI brain was normal. CT thoracic and lumbar spine was normal. Lab tests were within normal limits except Creatinine kinase levels were more than 300 (20 to 200u Normal)

Preoperative Preparation

High risk consent was taken in view of intraoperative hyperthermia, hyperkalemic cardiac arrest & post - operative ventilation if required. Prior to induction of anaesthesia, all the vaporizers were discontinued from the anaesthesia machine, fresh soda lime was filled into the soda lime canisters & breathing circuits were changed. Then the anesthesia machine was flushed at flow of 10L/min for 30min prior to remove any residual anaesthetic gases residue from prior use. It was ensured that there was absolutely no trace of inhalational anaesthetic in the anaesthesia workstation and circuit as shown by agent gas monitoring. Dantrolene was kept ready for malignant hyperthermia. ECG, SpO2, NIBP, capnography and temperature monitors were used for monitoring patient. Inj glycopyrolate 0.1mg I v, dexamethasone 4mg, inj fentanyl 50mcg was given child was induced with 80mg propofol, pre - oxygenated with 100% oxygen with bag mask ventilation and 5mg cisatracurium was given to facilitate endo tracheal intubation.5.5 south pole cuffed endotracheal tube was used. Intubation Cormack Lehane was grade 2. Anaesthesia was maintained with O2 +N2O+ Propofol infusion 100 to 200mcg/kg/min. Patient was given positive pressure ventilation & to maintain Etco2 30 to 35mm of Hg, Inj diclofenac was given in 50ml of NS IV. Intraoperative multiple episodes of hemodynamic fluctuation (systolic blood pressure from 80mm Hg to 130mm Hg & heart rate 70 to 130bpm) were managed by adjusting depth of anaesthesia by propofol infusion. ECG was normal intraoperative. Propofol infusion was stopped 10min before end of surgery & muscle paralysis was reversed after surgery. Child was extubated when he was fully awake with good respiratory effort. The child was kept in the intensive care unit for 48 hr where he made a successful recovery with no signs of rhabdomyolysis or hyperkalemia and shifted to ward for further care. He was discharged from the hospital on the 3rd postoperative day.

3. Discussion

The anaesthetic concern in a child with Duchenne's muscular dystrophy are difficult intubation, prolonged duration of neuromuscular blockade, possible need for postoperative ventilation, rhabdomyolysis hyperthermia and cardiac arrythmias when exposed to halogenated volatile anaesthetic agents & depolarizing muscle relaxant (2)

There are various reports of difficult intubation in patients with Duchenne's muscular dystrophy. (4.5In retrospective study by Muenster et al (6) difficult laryngoscopy was found in 4% of patients, the incidence was 7.5% in older children.

Obesity, large tongue, restricted mouth opening & limited mobility of cervical spine are thought to be causes of difficult intubation in this group of patients. Various studies have shown response of non - depolarizing muscle relaxant is abnormal. (5, 7) Delayed onset of action & prolonged effect have been seen after the use of non - depolarizing neuromuscular blocking agents. (5) PFT should be done to assess the risk of post - operative pulmonary complications & need for post op ventilation. TIVA based anaesthesia is considered safest in these patients. (6) N2O is considered safe in these patients. Precautions should be taken to ensure that patient is not exposed to inhalational agents during the surgery. The clinical course of Duchenne muscle dystrophy is severe & there is no causative therapy available. Some patients are on chronic corticosteroid therapy because this slows the progression of the disease. This disorder is characterized by progressive skeletal muscle weakness with an early onset in childhood. Muscle Re - organization with fatty infiltration & increase in fibrous tissue leads to loss of ambulation by 10 - year of age Perioperative morbidity is usually due to respiratory complications. Patients with vital capacities less than 30% of predicted appear to be at greatest risk and often require postoperative mechanical ventilation. (1) The anaesthetic management of these patients is complicated not only by muscle weakness but also by cardiac and pulmonary manifestations. An association with malignant hyperthermia has been suggested but is unproven. Succinylcholine should be avoided in patients with Duchenne's muscular dystrophy because of unpredictable triggering malignant hyperthermia. (10) TIVA based anaesthesia is considered safest in these patients. Intraoperative positioning may be complicated by kyphoscoliosis or by flexion contractures of the extremities or neck.

Although some patients exhibit a normal response to NDMBs, others may be very sensitive. (5, 7) Marked respiratory and circulatory depression may be seen with volatile anesthetics in patients with advanced disease, and regional or local anesthesia may be preferable in these patients.

4. Conclusion

Comprehensive preoperative evaluation to discern functional status of each particular patient, anticipate perioperative risk and provide specific management in muscular dystrophy patients is necessary to reduce morbidity or mortality associated with disease and anaesthetic management. Avoidance of anaesthetic agents which may trigger rhabdomyolysis and severe hyperkalemia are the keys to successful anaesthesia outcome in patients with Duchenne muscular dystrophy.

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