

# Anaesthetic Management in Duchenne Muscular Dystrophy Patient with TIVA Using Combination of Propofol and Dexmedetomidine Complimented with USG Guided ESPB Block: A Case Report

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## ABSTRACT

Muscular dystrophies are a group of genetic diseases which cause progressive degeneration of skeletal muscle along with weakness. They are characterized by an impaired synthesis or regeneration of contractile protein. Patients with muscular dystrophies possess high preoperative risk with significant anaesthetic implications (hyperkalaemia, rhabdomyolysis, cardiovascular instability, sudden death, etc.). We herein describe the anaesthetic management of a 31yr old male who was a known case of Duchenne muscular dystrophy posted for right percutaneous nephrolithotomy (PCNL). The patient was induced and maintained with TIVA and using nitrous oxide as the only inhalational agent supplemented with USG guided Erector spinae plane block (ESPB) for post-operative analgesia. A thorough preoperative evaluation and multidisciplinary approach is essential for perioperative management of such cases.

Duchenne muscular dystrophy (DMD) most commonly affects the paediatric age group about 1 in 3500 live male births. DMD is caused by mutations in the dystrophin gene located on chromosome Xp21 resulting in an abnormal form or very low concentration of dystrophin [1]. The muscular tissue here, is replaced by fatty infiltrates and fibrous tissue causing degeneration of skeletal muscle. There is progressive proximal muscle weakness that begins in childhood [2]. These patients are usually wheelchair bound by adolescence and develop cardiac or pulmonary manifestations either towards late 20's or early 30's. In such patients, anaesthetic challenges are faced when they are posted for elective/emergency surgery, as there is a risk of rhabdomyolysis, leading to hyperkalaemia, increased muscle weakness, cardiac arrest, malignant arrhythmias triggered by anaesthetic agents, and exacerbation of respiratory failure [3]. This subset of patients present with macroglossia and restricted mobility

of mandible and cervical spine resulting in a difficult airway. We present a patient with DMD posted for right PCNL who underwent successful anaesthetic management.

## Case Report

A 31-year-old male, weighing 70 kg, presented to the anaesthesia OPD with complaints of pain in right flank radiating to back for 20 days with history of vomiting 2-3 episodes for 5 days, dyspnoea on exertion (DOE) Grade 2 and hoarseness of voice. He was a diagnosed case of DMD since childhood. He had progressive weakness of bilateral lower limbs followed by bilateral upper limb involvement. Patient had history of left URS in 2016 under spinal anaesthesia and bone marrow transplant in 2018 under GA.

On examination, the patient was in sitting position. The power of both upper and lower limbs was 4/5 with waddling gait. Rest of the systems within normal limits.

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Chest radiograph showed no abnormality. ECG showed sinus tachycardia. Airway examination revealed mallampati grade 2 with neck flexion and extension adequate. Breath holding time was 30s. All routine investigations were within normal limits. CT urography showed right moderate hydronephrosis with proximal hydroureter due to mild ureteric calculus (10x7x16mm). Physician advised 2Decho and it was normal, cardiologist opinion regarding cardiac status was sought and patient was posted for elective surgery after obtaining informed high-risk consent.

After confirming NBM status (eight hours), ICU bed with ventilatory support and blood/blood products, patient was taken inside the O.T. Pulse oximeter, non-invasive blood pressure, ECG leads were attached and 18 G intravenous (IV) access was secured. Before induction, vaporizers were dismantled from the anaesthesia machine and fresh soda lime canister along with new breathing circuits were attached. For removal of any residual anaesthetic gases from prior use, the anaesthesia machine was flushed at a fresh gas flow of 8 l/min for 20 min [5]. It was ensured that no inhalational anaesthetic remained inside the anaesthesia machine which was confirmed by anaesthesia gas analyser. Difficult airway cart with McCoy laryngoscope, video laryngoscope, Airtraq laryngoscope and endotracheal tubes of all sizes ranging from 7.0 to 8.5 were kept ready. All emergency equipment and drugs were checked and kept ready, OT temperature was kept warm and warm blankets were also kept ready.

Patient was premedicated with inj. pantoprazole 40 mg IV given prophylactically to prevent aspiration. Premedication was done with inj. glycopyrrolate 0.2mg, inj. ondansetron 4mg following which inj. dexmedetomidine was administered in a loading dose of 0.6 µg/kg was given IV. After preoxygenation with 100% oxygen, inj. fentanyl 100mcg was given as analgesic. Intravenous induction was done by inj propofol 100 mg IV, rocuronium bromide 70 mg IV and rapid sequence induction guidelines were followed securing airway with ETT 8.5mm and position was confirmed with auscultation and capnography. Maintenance of anaesthesia was achieved with intravenous propofol infusion 5mg/kg/hr and dexmedetomidine infusion 0.5mcg/kg/hr with O<sub>2</sub>: N<sub>2</sub>O= 50:50. The ventilator was set with a respiratory frequency 12 /minute, tidal volume of 450 mL and inspiratory/expiratory time ratio 1: 2 to maintain etCO<sub>2</sub> between 35-40 mmHg. Patient was then given prone position taking care of all pressure points and the airway pressure was also monitored. Antifibrinolytic therapy with inj. tranexamic acid 1gm IV was administered before surgical incision. Blood loss was minimal (150ml) and the calculated total amount of fluid was replaced with potassium free crystalloids (1000ml NS and 500ml DNS) through a fluid warmer.

Patient was hemodynamically stable throughout the procedure which lasted for 3h. For postoperative analgesia USG guided ESPB was given. Propofol and dexmedetomidine infusion was stopped 15 min prior to

the end of surgery and neuromuscular reversal was done with inj neostigmine 2.5mg IV and inj glycopyrrolate 0.4mg I.V. Patient was extubated after he was fully awake with good respiratory efforts. The patient was transferred to intensive care unit with stable vitals for post-operative care.

## Discussion

Duchenne muscular dystrophy (DMD) possesses multiple challenges for anaesthetic care. Both invasive and non-invasive procedures requiring either sedation or general anaesthesia are necessary for these affected patients. The major anaesthetic challenges in the management of patients with DMD is the use of volatile (halogenated) inhalational agents as well as depolarizing relaxants because of the potential for rhabdomyolysis and severe hyperkalaemia [6-7].

Administration of anaesthesia in such patients is associated with numerous challenges and can result in varied spectrum of metabolic and clinical complications such as rhabdomyolysis, metabolic derangements, malignant hyperthermia, cardiac arrhythmia and even death [3]. Due to these challenges, a multidisciplinary team is required for thorough pre-operative evaluation, intraoperative management and post-operative care. Conduction defects and cardiomyopathy are common in these patients because of degeneration of cardiac muscle fibres. Baseline ECG and echocardiography should be done preoperatively. It is proven that patients with left ventricular ejection fraction of less than 0.5 stand a higher risk of perioperative complications [3].

Antifibrinolytic therapy and blood sparing techniques should be preferred so as to decrease blood loss and resultant hypothermia [4]. Hypothermia with shivering can induce a myotonic contracture [8]. Vigilant temperature monitoring and warming strategies with either warm fluids, forced air warmers or electric blankets to prevent hypothermia should be used [8]. Potassium free crystalloids should be preferred and hence, glucose solutions are recommended so as to reduce the risk of rhabdomyolysis and acute hyperkalaemia [3]. Due to large tongue, weak dilator muscles of the upper airway, the inability to maintain patency of upper airway owing to limited mobility of mandible and cervical spine and hypoxemia due to reduced functional residual capacity, these patients are often prone to upper airway obstruction [9]. Hence, a difficult airway cart with newer gadgets should also be kept ready.

Furthermore, patients with muscular dystrophies are at a greater risk of aspiration as there is weakness of the pharyngeal muscles and a reduced gastric emptying time. Hence, aspiration prophylaxis and rapid sequence induction is recommended in these patients. However, the use of depolarizing muscle relaxant succinylcholine is contraindicated as it causes hyperkalaemia, rhabdomyolysis, exaggerated contracture and masseter spasm [10]. Therefore, non-depolarizing muscle

relaxants with a short recovery index like rocuronium is recommended [11]. Rocuronium is a non-depolarizing neuromuscular blocking agent with a rapid onset of action and is preferred for routine or rapid sequence intubation in both adult and paediatric patients [12].

Inhalational anaesthetic agents should be avoided as they cause rhabdomyolysis and may precipitate malignant hyperthermia. Gronert et al. in his study, described two two-year old boys who underwent muscle biopsies that established the histopathological diagnosis of Becker's dystrophy in one, and Duchenne dystrophy in the other. Concomitant contracture testing with caffeine or halothane was normal for malignant hyperthermia (MH). He suggested that acute hypermetabolism or acute rhabdomyolysis during anaesthesia, in patients with these disorders, is related to the X-linked myopathy and its associated muscle deterioration, rather than to the autosomal dominant MH [13]. However, Muenster et al. reviewed anaesthetic management of 232 cases of muscular dystrophies undergoing various orthopaedic surgeries and concluded that nitrous oxide in oxygen was used safely in 135 cases with no adverse events [14].

Multiple previous studies have shown unpredictable response to non-depolarizing muscle relaxants in these patients, with delayed onset and prolonged duration of action. For neuromuscular monitoring a peripheral nerve stimulator can be used but the electrical stimulus can itself cause myotonia [8]. Hence rocuronium was used only in the beginning to facilitate intubation [15-16]. For PCNL both spinal and general anaesthesia are effective techniques. However, there have been reported cases of myotonic contractures following incomplete central neuraxial blockade [17]. In addition, spinal anaesthesia can also be challenging due to the spinal abnormalities in these patients [4]. Due to these reasons, total intravenous anaesthesia with combination of two or more sedative agents like propofol, dexmedetomidine and ketamine is the recommended option for maintenance.

Opioids can also be used for maintenance of anaesthesia but as DMD patients are more sensitive to them they are avoided. NSAIDs may trigger rhabdomyolysis. However, paracetamol is considered to be safe for providing analgesia. Bennun et al; found that propofol at an induction dose of 2.5 mg/kg followed by combination of propofol infusion at 6mg/kg/hour, fentanyl, and atracurium with nitrous oxide in oxygen appears to be a suitable anaesthetic technique for young myotonic dystrophy patients undergoing peripheral surgery [18].

Rozmiarek et al; reported a case with successful use of dexmedetomidine and ketamine combination to provide sedation and analgesia in a 21-year-old patient with DMD undergoing bone marrow aspiration and biopsy [19]. Whereas, Kako et al; used 2 different doses of dexmedetomidine for sedation during muscle biopsy in patients with DMD: 1.0 or 0.5 µg/kg was administered as a loading dose over 3 minutes followed by a continuous infusion of 1.0 or 0.5 µg/kg/hour [20].

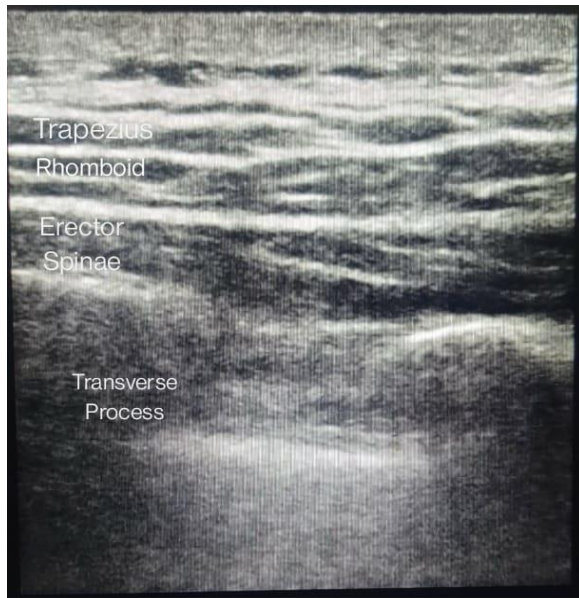
Similar to our study, Gaszynski reported the successful use of dexmedetomidine and propofol for maintenance of TIVA following RSI using rocuronium in a 21-year-old male patient with myotonic dystrophy posted for laparoscopic cholecystectomy. Inj propofol 250mg followed by continuous infusion, inj rocuronium 20mg and inj dexmedetomidine 0.6 µg/kg over 10 minutes followed by continuous infusion of dose of 0.2 µg/kg/hour was used without any complications [21].

For reversal of neuromuscular blockade neostigmine or sugammadex are the two agents which can be used. Sugammadex is a newer selective binding agent for reversal with a high affinity for rocuronium. In adult patients, according to a moderate-quality RCT, a higher number of patients treated with 4 mg/kg sugammadex experienced hypoxemia within 24 hours after treatment as compared to those treated with 40 mcg/kg neostigmine with 10 mcg/kg atropine [22]. It is also expensive and not easily available in India. Therefore, we preferred neostigmine for reversal.

Extubation should be done only when the patient is completely awake and there is no residual paralysis following reversal of blockade. Post-operative monitoring in the intensive care unit is necessary for delayed complications like cardiovascular instability, arrhythmias, respiratory depression and rhabdomyolysis [23].

Optimal post-operative pain management is also challenging. Opioids though considered to be safe may cause respiratory depression, gastroparesis, acid reflux and aspiration, hence should be avoided [24-25]. Peripheral nerve blocks are considered to be safe and can be used for post-operative analgesia. Acute pain in the postoperative period after PCNL is mainly due to visceral pain originating from the kidneys and ureters (T10-L2) and somatic pain from the incision site (T8-12). Erector Spinae plane block (ESPB) is a simple paraspinal, fascial block targeting the ventral, dorsal rami and rami communicantes of spinal nerves [26]. Ibrahim et al. conducted a study on 56 patients undergoing elective PCNL and concluded that the group in which ESPB was given preoperatively had better post-operative analgesia and lesser opioid consumption [27].

Hence, we gave USG guided ESPB with Inj. Ropivacaine 0.375% for post-operative analgesia. Our patient was pain free for eight hours with a VAS score of less than four. Adequate post-operative analgesia facilitates early recovery of patients.

**Figure 1- USG appearance of Erector Spinae Block**

Our patient did not give any history of awareness during the procedure. Dexmedetomidine, a highly selective  $\alpha$ -2 agonist, due to sedative, analgesic, sympatholytic, narcotic and volatile agent sparing property makes it a suitable choice as an anaesthetic adjuvant for TIVA especially in DMD. DMD is a challenge for anaesthesiologists with respect to airway, metabolic and clinical complications related to the disease itself and anaesthetic agents. There is limited literature about TIVA plus USG guided ESPB to provide adequate surgical anaesthesia and post-operative analgesia for PCNL especially in cases with muscular dystrophy.

### Conclusion

Comprehensive preoperative evaluation to discern functional status of each particular patient and anticipate perioperative risk and provide specific management in DMD patients is necessary to reduce morbidity or mortality associated with disease and anaesthetic management. We suggest combination of propofol and dexmedetomidine (for maintenance) in TIVA technique complimented with USG guided ESPB for postoperative analgesia for safe outcome in DMD cases.

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