

'Please Don't Intubate Me': Managing Laparoscopic Surgery on a Duchenne Muscular Dystrophy Patient with Severe Restrictive Pulmonary Disease

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INTRODUCTION

- Duchenne muscular dystrophy (DMD) is a hereditary degenerative disorder characterized by progressive muscle weakness, respiratory insufficiency and cardiomyopathy leading to significant perioperative challenges.
- Patient request to avoid intubation associated with general anesthesia poses significant challenges for anesthetic management and requires employment of unique strategies.

CASE PRESENTATION

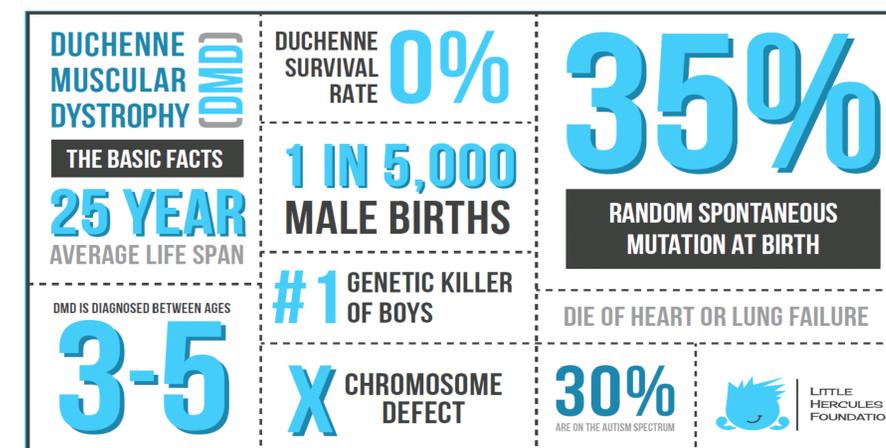
- A 17-year-old 40 kg male with DMD and severe restrictive respiratory failure with a FVC of 10% requiring continuous nasal BiPAP presented for laparoscopic-assisted gastrostomy tube placement for malnourishment.**
- Patient refused procedure unless performed without intubation due to high risk of prolonged postoperative intubation and permanent tracheostomy dependence.
- TTE demonstrated EF of 60%. Previous posterior spinal fusion from T1 – L4 precluded an epidural.
- Patient agreed to a combination of sedation and regional techniques and to abort procedure if not tolerated.
- Intraoperative management began with midazolam 2 mg IV and dexmedetomidine infusion with BiPAP in place.
- Bilateral US guided rectus sheath and unilateral left transversus abdominis plane block were performed.
- Lidocaine was infiltrated into the incision sites to supplement.
- Prior to insufflation, a remifentanyl infusion was started and ketamine 10mg IV given. During the insufflation, patient asks, "have we started the surgery yet?"
- His respiratory and hemodynamics remained stable throughout.
- In recovery, he complained of 8/10 pain at the gastrostomy site which responded to a small dose of IV opioid.
- He was discharged home the next day on his baseline BiPAP support.

DMD RESPIRATORY STAGES		
Ambulatory Stage	Early Non-ambulatory Stage	Late Non-ambulatory Stage
Assessments		
Once yearly: FVC	Twice yearly: FVC, MIP/MEP, PCF, SpO ₂ , p _a CO ₂ /ptcCO ₂	
Sleep study with capnography for signs and symptoms of obstructive sleep apnoea or sleep-disordered breathing		
Interventions		
Immunisation with pneumococcal vaccines and yearly inactivated influenza vaccine		
Lung volume recruitment with FVC <60% predicted		
Assisted coughing when FVC <50% predicted, PCF <270 L/min, or MEP <60 cm H ₂ O		
Nocturnal assisted ventilation with back-up rate of breathing (non-invasive preferred when there are signs or symptoms of sleep hypoventilation or other sleep-disordered breathing, abnormal sleep study, FVC <50% predicted, MIP <60 cm H ₂ O, or awake baseline SpO ₂ <95% or pCO ₂ >45 mm Hg		
Addition of assisted daytime ventilation when, despite nocturnal ventilation, daytime SpO ₂ <95%, pCO ₂ >45 mm Hg, or symptoms of awake dyspnoea are present		

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DISCUSSION

- DMD is the most common form (~1 in 3500 males) of inherited muscle disease with mutations in the dystrophin protein leading to total absence of dystrophin resulting in progressive muscle weakness and cardiomyopathy.
- Symptoms appear at 2-3 years of age, with nearly all patients losing independent ambulation by age 5.
- Respiratory insufficiency and cardiomyopathy affect 90% by age 30, followed by death in the third decade.
- American College of Chest Physicians consensus statement in 2009 on management of patients with DMD undergoing General Anesthesia (GA) or sedation advised the avoidance of GA in favor of neuraxial or regional anesthesia techniques whenever possible due to limited cardiac function and chronic respiratory depression, as well as the facts that inhalational anesthetics have been implicated in causing rhabdomyolysis and succinylcholine is absolutely contraindicated for risk of rhabdomyolysis and hyperkalemic cardiac arrest.



CONCLUSIONS

- Patients with DMD are increasingly living longer they present with challenging perioperative risks of compromised cardiopulmonary and airway management.
- Perioperative management of advanced DMD patients requires proactive collaboration of multiple professional disciplines to ensure the best possible outcome, as well as serious consideration to the avoidance of general anesthesia with invasive airway where post-operative risk of inability to extubated is high.

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