

Anesthetic Management of a Patient with Muscular Dystrophy and Scoliosis Undergoing Emergency Open Appendectomy: A Case Report

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Abstract

Patients with muscular dystrophy combined with scoliosis present significant anesthetic challenges due to risks of respiratory compromise, cardiac dysfunction, altered responses to anesthetic agents, and susceptibility to rhabdomyolysis or malignant hyperthermia. Thorough pre-operative evaluation—including pulmonary and cardiac assessment—is essential. This case describes the successful management of a 35-year-old male with muscular dystrophy and severe scoliosis undergoing emergency open appendectomy under general anesthesia using total intravenous anesthesia with propofol, short-acting opioid analgesia, and avoidance of neuromuscular blockers for intubation. The procedure and postoperative course were uneventful. This case highlights key anesthetic considerations for patients with neuromuscular disorders requiring emergency abdominal surgery.

Keywords: Muscular dystrophy; scoliosis; anesthesia; total intravenous anesthesia; propofol; neuromuscular disorders

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Introduction

Muscular dystrophies are a heterogeneous group of genetic disorders caused by defects affecting muscle fiber integrity and function. These disorders vary in clinical presentation and genetic basis but commonly lead to progressive skeletal muscle weakness and multisystem involvement, including cardiomyopathy, respiratory dysfunction, and risk of malignant arrhythmias and rhabdomyolysis during anesthesia.¹

Scoliosis further compounds anesthetic challenges by contributing to restrictive lung physiology, reduced lung compliance, ventilation–perfusion mismatch, and potential pulmonary hypertension and right-sided heart strain.² Patients with both neuromuscular disease and spinal deformity require individualized anesthetic planning.

Although regional anesthesia avoids many anesthetic risks, its use is limited in severe scoliosis, especially during emergency abdominal surgery. Thus, a carefully tailored general anesthesia plan is often required. We present the successful management of such a patient undergoing emergency open appendectomy.

Case History

A 35-year-old male weighing 40 kg presented for emergency open appendectomy. He had a history of muscular dystrophy diagnosed at 8 years of age, leading to progressive weakness of all limbs and wheelchair dependence by age 10. He also had severe scoliosis and global developmental delay with cognitive impairment below age-appropriate norms. No family history of neuromuscular disorders was reported.

Examination revealed marked wasting of bilateral upper and lower limbs. Airway assessment demonstrated no predictors of difficult intubation. Vital signs were normal. Laboratory investigations, including complete blood count, metabolic profile, and urinalysis, were within normal limits. ECG showed sinus rhythm with right axis deviation. Echocardiography revealed mild tricuspid regurgitation (peak gradient 24 mmHg), trace aortic regurgitation, and preserved left ventricular systolic function (LVEF 60%).

The patient and family were counseled regarding perioperative risks, and high-risk informed consent was obtained. An ICU bed was reserved pre-operatively.

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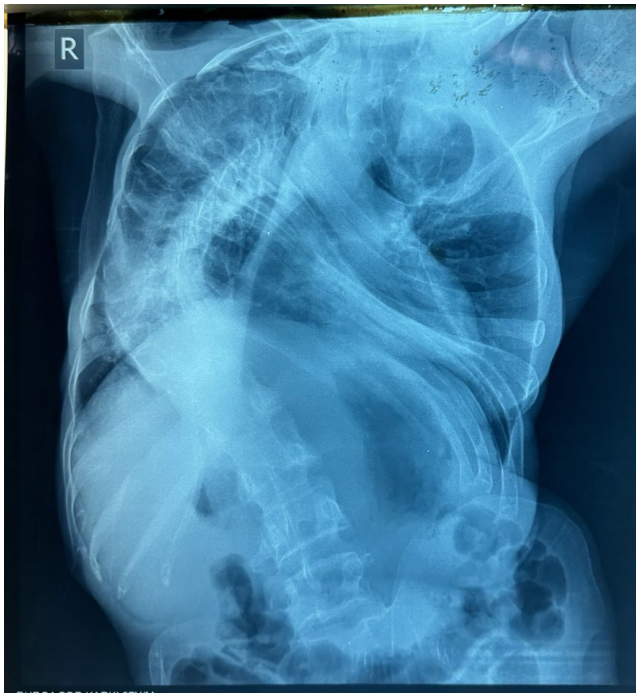


Figure 1: Preoperative chest X-ray.



Figure 2: Clinical appearance of the patient.

Intraoperative Management

The anesthesia workstation was prepared with precaution: vaporizers were removed, the breathing circuit was changed, and the machine was flushed with 10 L/min oxygen for 20 minutes. Standard ASA monitoring along with temperature and capnography was applied.

A 20-gauge intravenous cannula was secured, and the patient was positioned with adequate padding to prevent pressure injuries due to muscle wasting.

Induction was performed with fentanyl 40 µg and propofol 100 mg. Laryngoscopy with a Macintosh blade was uneventful, and tracheal intubation was performed with a 6.5-mm cuffed endotracheal tube. Cormack–Lehane grade was IIa. Mask ventilation and intubation conditions were adequate without neuromuscular blockade. Hemodynamics remained stable.

Anesthesia was maintained using propofol infusion at 50 µg/kg/min. Vecuronium 3 mg was given for intraoperative muscle relaxation. Capnography, temperature, and ECG showed no signs of malignant hyperthermia, arrhythmias, or hypercarbia. Surgical duration was 1 hour.

At the end of surgery, neuromuscular recovery was facilitated with neostigmine 1.5 mg and glycopyrrolate 0.3 mg, with adequate return of spontaneous ventilation confirmed clinically. The patient was extubated once tidal volume exceeded >6 mL/kg, airway reflexes returned, and he obeyed verbal commands.

Postoperative Course

The patient was monitored in the postoperative care unit for 48 hours, with no respiratory compromise or hemodynamic instability. Analgesia was provided with IV paracetamol 750 mg three times daily and IV ketorolac 15 mg as needed. He was shifted to the ward on postoperative day 3 and discharged on day 4.

Discussion

Muscular dystrophies involve progressive degeneration of muscle fibers due to genetic defects. These patients are at increased risk of perioperative complications including malignant hyperthermia-like reactions, rhabdomyolysis, cardiac arrhythmias, and postoperative respiratory insufficiency.¹ Scoliosis further impairs respiratory mechanics, decreases functional residual capacity, and may increase the risk of pulmonary hypertension and right-heart dysfunction.²

Anesthetic Considerations

Regional anesthesia is generally preferred where feasible because it avoids risks associated with inhalational anesthetics and neuromuscular blockers. However, in emergency abdominal surgery with significant spinal deformity, regional anesthesia may be technically difficult and insufficient.

Total intravenous anesthesia (TIVA) with propofol is considered safer for patients with muscular dystrophy due to reduced risk of rhabdomyolysis compared with volatile anesthetics.³ Opioid sensitivity in neuromuscular disorders requires cautious dosing; therefore, a short-acting opioid (fentanyl) was selected.⁴

Succinylcholine is contraindicated because it may trigger hyperkalemia and rhabdomyolysis. Non-depolarizing neuromuscular blockers may have prolonged effects due to abnormal muscle physiology and are best avoided unless neuromuscular monitoring is available.⁵

Intraoperative Concerns

Continuous temperature and end-tidal CO₂ monitoring are essential to detect early signs of malignant hyperthermia.⁶ In this case, no abnormalities were observed.

Careful patient positioning is crucial due to muscle wasting and skeletal deformity, reducing the risk of nerve compression or pressure sores.

Limitations

A definitive subtype of muscular dystrophy could not be confirmed due to the unavailability of muscle biopsy or genetic testing. Neuromuscular and depth of anesthesia monitoring were unavailable, which would have improved drug titration and safety.

Comparison with Literature

Previous reports describe successful regional anesthesia for abdominal and gynecologic surgeries in muscular dystrophy patients.^{3,4} However, these were elective cases with more favorable spinal anatomy. In contrast, our patient required emergency laparotomy where regional anesthesia was unsuitable, consistent with other reports advocating TIVA-based general anesthesia in similar scenarios.⁵

Conclusion

General anesthesia in patients with muscular dystrophy and scoliosis poses significant challenges. However, with careful preoperative assessment, avoidance of triggering agents, use of short-acting opioids, and TIVA with propofol, safe anesthetic management is achievable. This case reinforces the importance of individualized planning and vigilant perioperative monitoring in neuromuscular disorders.

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Conflict of Interest

None declared.

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Patient Consent for Publication

Written informed consent for publication was obtained from the patient and guardian.

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