Research Square

Preprints are preliminary reports that have not undergone peer review.
They should not be considered conclusive, used to inform clinical practice,
or referenced by the media as validated information.

Perioperative Anesthetic Management of a Child with Progressive Muscular Dystrophy Undergoing Preauricular Fistula Resection: A Case Report

Zhiwei Zhang
Graduate School of Hebei North University
Menghan Ning
the First Affiliated Hospital of Hebei North University
Congna Zi
zicongna868@163.com

the First Affiliated Hospital of Hebei North University

Case Report

Keywords: Progressive muscular dystrophy, Duchenne muscular dystrophy, Preauricular fistula, Total intravenous anesthesia, Perioperative management

Posted Date: June 17th, 2026

DOI: https://doi.org/10.21203/rs.3.rs-9673876/v1

License: This work is licensed under a Creative Commons Attribution 4.0 International License.
Read Full License

Additional Declarations: No competing interests reported.

Abstract

Background: Duchenne muscular dystrophy (DMD) poses high anesthetic risks due to dystrophin deficiency. Reports on perioperative management for DMD children undergoing non-orthopedic surgery are limited.

Case presentation: A 4-year-old boy with DMD underwent preauricular fistula resection. Total intravenous anesthesia with propofol and remifentanil was used, avoiding volatile agents. Low tidal volume kept peak airway pressure at 12 cmH_2O. Surgery lasted 35 min. Spontaneous breathing resumed at 6 min; extubation was uneventful. No complications occurred.

Conclusions: TIVA with propofol and remifentanil, along with low airway pressure, active warming, and early extubation, enabled safe management for this case. This provides a clinical reference for similar patients.

Background

Progressive muscular dystrophy (PMD) comprises rare hereditary muscle disorders; Duchenne muscular dystrophy (DMD) has the highest incidence. DMD results from *DMD* gene mutations on the X chromosome, causing complete dystrophin absence. The disease typically presents in early childhood with progressive skeletal muscle atrophy and weakness, along with markedly elevated creatine kinase [1]. Children with DMD often have concurrent myocardial and respiratory dysfunction and exhibit abnormal responses to depolarizing muscle relaxants and volatile anesthetics, which may trigger rhabdomyolysis, hyperkalemia, or cardiac arrest [2]. Reports on non-orthopedic head and neck surgery in children with DMD are limited. This case shares our perioperative strategy for a child with DMD and infected preauricular fistula, aiming to provide a clinical reference for similar rare disease patients.

Case presentation

A 4-year-old boy (96 cm, 15 kg) was admitted for recurrent left preauricular redness, swelling, and pain for 2 years, aggravated for 7 days. A pinhead-sized sinus tract had been present since birth. Six months earlier, elevated transaminases (ALT 266.8 U/L, AST 179.3 U/L) were found; after excluding liver diseases, genetic testing confirmed DMD. He had received intermittent hepatoprotective agents but no glucocorticoids. No prior surgery or allergies. Physical exam: left preauricular fluctuating swelling; muscle strength grade V, Gowers' sign negative. Laboratory: CK 19,888 U/L, LDH 1,156.8 U/L, CK-MB 294.9 IU/L, AST 264 U/L, ALT 281 U/L. ECG: sinus tachycardia (114 bpm). Echocardiography and pulmonary function normal. Diagnoses: left preauricular fistula with infection and DMD. Written informed consent was obtained.

After entering the operating room, monitoring included ECG, noninvasive blood pressure, pulse oximetry, nasopharyngeal temperature, and BIS. Induction: fentanyl 50 μg, propofol 35 mg, rocuronium 5 mg. A 4.5-mm endotracheal tube was inserted. Maintenance: TIVA with propofol 10 mg/(kg·h) and remifentanil

0.25 μg/(kg·min); no volatile anesthetics. Ventilation: volume-controlled, tidal volume 120 mL, rate 18 bpm, PEEP 0 cmH_2O, FiO_2 50%; peak airway pressure 12 cmH_2O. Active warming maintained temperature 36.5–37.0°C. Surgery lasted 35 min with ~ 1 mL blood loss. No myotonia, rhabdomyolysis, hyperkalemia, or arrhythmias occurred. Anesthetics stopped 5 min before end; spontaneous breathing resumed at 6 min, and the patient was extubated uneventfully. Postoperative recovery was smooth; no respiratory depression. The child was discharged on day 3. At 1-month follow-up, no recurrence and good healing.

Discussion and Conclusions

DMD is the most common and severe PMD subtype, affecting ~ 1/3,300–5,000 live male births [3]. It results from X-linked *DMD* mutations causing complete dystrophin deficiency, which destabilizes the sarcolemma [4]. Without dystrophin, contraction-induced injury leads to progressive fibrosis and muscle weakness [5]. Pathological changes also affect the myocardium and respiratory muscles. Cardiomyocyte instability permits excessive calcium influx, triggering myocyte death and fibrosis, ultimately leading to dilated cardiomyopathy [6]. Although our patient had a normal echocardiogram at age 4, his markedly elevated CK (19,888 U/L) indicated widespread sarcolemmal damage typical of early-stage DMD [7]. Respiratory involvement from diaphragmatic and intercostal weakness leads to restrictive lung disease and hypoventilation. Even young children may have sleep-disordered breathing with CO_2 retention [8].

DMD patients are at risk of life-threatening reactions to specific anesthetics. Depolarizing muscle relaxants like succinylcholine can trigger massive potassium release, causing hyperkalemia and cardiac arrest [9]. Volatile anesthetics have been associated with rhabdomyolysis, hyperkalemia, and a malignant hyperthermia-like syndrome, likely due to dystrophin deficiency-related calcium dysregulation and oxidative stress [10]. Thus, trigger agents are absolutely contraindicated. Current guidelines strongly recommend TIVA for DMD patients [11]. We used propofol-remifentanil TIVA without volatile agents. The patient remained hemodynamically stable with no myotonia or arrhythmias. Rocuronium 5 mg was given for induction, and the child recovered spontaneous muscle strength rapidly, with no residual block.

Even for minor procedures like preauricular fistula resection, several intraoperative issues require attention. First, weak respiratory muscles mandate low tidal volumes to maintain low peak airway pressures [7]. We kept peak pressure at 12 cmH_2O. Second, temperature homeostasis is critical. DMD patients are prone to both hypothermia and hyperthermia. Active warming with a forced-air blanket helped maintain nasopharyngeal temperature at 36.5–37.0°C [12]. Third, careful head positioning is needed to avoid neck injury. Close communication between anesthesiologist and surgeon is essential. We adhered to a “minor surgery, major management” philosophy.

Postoperative care is critical. Respiratory muscle weakness predisposes to atelectasis and retained secretions. Our patient resumed spontaneous breathing at 6 minutes and was extubated early, with no hypoventilation. For DMD patients with restrictive lung disease, postoperative non-invasive ventilation should be considered [13]. Pain management must avoid respiratory depression: opioids minimized. We

used NSAIDs as the primary analgesic, avoiding high doses that could impair renal function. Regional blocks (e.g., auricular nerve block) are an alternative.

This case has several distinctive aspects compared with previously reported DMD anesthesia experiences[14][15]. First, our patient was only 4 years old, whereas most published cases involve school-aged children (7–20 years), implying lower organ reserve and higher risk. Second, the child had been diagnosed for only six months and had not received standardized glucocorticoid therapy. Third, preauricular fistula resection imposes specific demands on head positioning and airway sharing. Core lessons: (1) multidisciplinary preoperative assessment is essential; (2) TIVA with propofol and remifentanil, avoiding all trigger agents, is safe and effective; (3) low peak airway pressure, active temperature management, and reduced doses of non-depolarizing relaxants contribute to stability; (4) early extubation and vigilant postoperative monitoring promote uneventful recovery. In conclusion, with careful TIVA-based planning, DMD children can safely undergo minor non-orthopedic surgeries.

Abbreviations

ALT
Alanine aminotransferase
AST
Aspartate aminotransferase
BIS
Bispectral index
CK
Creatine kinase
CK-MB
Creatine kinase myocardial band
DMD
Duchenne muscular dystrophy
ECG
Electrocardiogram
FiO_2
Fraction of inspired oxygen
LDH
Lactate dehydrogenase
NSAIDs
Non-steroidal anti-inflammatory drugs
PEEP
Positive end-expiratory pressure
PMD
Progressive muscular dystrophy
TIVA

Total intravenous anesthesia

Declarations

Ethics approval and consent to participate:

This case report was part of a study on perioperative anesthesia management for patients with rare diseases (project title: “Perioperative Anesthesia Management for Patients with High-Risk Special Diseases Undergoing Surgery”). The study protocol (version 1.0) was approved by the Medical Ethics Committee of the First Affiliated Hospital of Hebei North University (ethics approval number: [K2026137];approval date: [26 February 2026]). All procedures were performed in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki Declaration and its later amendments.

Consent for publication:

Written informed consent for publication of this case report and any accompanying clinical data (including clinical history, physical examination findings, laboratory results, and treatment details) was obtained from the patient’s parent.

Competing interests:

The authors declare that they have no competing interests.

Funding:

Not applicable.

Author Contribution

Z.Z. (Zhiwei Zhang): conceptualization, data collection, writing – original draft. M.N. (Menghan Ning): clinical data acquisition, writing – review & editing. C.Z. (Congna Zi): conceptualization, case supervision, manuscript revision, corresponding author. All authors read and approved the final manuscript.

Acknowledgements:

Not applicable.

Availability of data and materials:

Not applicable.

Declaration of conflicts of interest:

None declared.

References

1. Kemper AR, Lam WKK, Ojodu J, Jones E, Smith HS, Comeau AM, et al. Evidence regarding Duchenne muscular dystrophy newborn screening. Pediatrics. 2025. 10.1542/peds.2025-073192.
2. Lian X, Jing Y, Luo T, Guo Y, Lin Y. Pharmacological interventions for the management of anesthesia and sedation in patients with Duchenne muscular dystrophy: a systematic review and meta-analysis. Front Med (Lausanne). 2025;12:1497538.
3. Crisafulli S, Sultana J, Fontana A, Salvo F, Messina S, Trifirò G. Global epidemiology of Duchenne muscular dystrophy: an updated systematic review and meta-analysis. Orphanet J Rare Dis. 2020;15(1):141.
4. Boyer O, Butler-Browne G, Chinoy H, Cossu G, Galli F, Lilleker JB, et al. Myogenic cell transplantation in genetic and acquired diseases of skeletal muscle. Front Genet. 2021;12:702547.
5. Farini A. Special issue: pathophysiology and therapeutic perspectives in DMD: the well-defined role of the immune system. Biomedicines. 2021;9(12):1911.
6. Carraro L, Iosca A, Dainesi MI, Fusco S, Chieffo DPR, Moriconi F, et al. Cognitive function in DMD carriers: personal case series and literature review. Acta Myol. 2023;42(2–3):53–9.
7. Zhang X, Wang HP, Feng XX. Research progress on clinical characteristics of different subtypes of progressive muscular dystrophy. Mod Clin Med. 2024;50(5):372–4. (in Chinese).
8. Prigent H. Respiratory care in Duchenne muscular dystrophy. Archives de Pédiatrie. 2025; 32(7): 7S25-7S31.
9. Radkowski P, Oniszczuk H, Opolska J, Podlińska I, Pawluczuk M, Onichimowski D. A review of muscle relaxants in anesthesia in patients with neuromuscular disorders including Guillain-Barré syndrome, myasthenia gravis, Duchenne muscular dystrophy, Charcot-Marie-Tooth disease, and inflammatory myopathies. Med Sci Monit. 2024;30:e945675.
10. Bez Batti Angulski A, Hosny N, Cohen H, Martin AA, Hahn D, Bauer J, et al. Duchenne muscular dystrophy: disease mechanism and therapeutic strategies. Front Physiol. 2023;14:1183101.
11. van den Bersselaar LR, Heytens L, Silva HCA, Reimann J, Tasca G, Díaz-Cambronero Ó, et al. European Neuromuscular Centre consensus statement on anaesthesia in patients with neuromuscular disorders. Eur J Neurol. 2022;29(12):3486–507.
12. van den Bersselaar LR, Snoeck MMJ, Gubbels M, Riazi S, Kamsteeg EJ, Jungbluth H, Voermans NC. Anaesthesia and neuromuscular disorders: what a neurologist needs to know. Pract Neurol. 2021;21(1):12–24.

13. Wahlgren L, Nordström S, Kroksmark AK, Tulinius M, Lindberg C, Sofou K. Living with Duchenne muscular dystrophy as an adult: Motor function, ventilatory assistance and health-related quality of life. Neuromuscul Disord. 2026: 106409.
14. Tsirikos AI, Roberts SB. The Development of Spinal Deformity in Patients with Duchenne Muscular Dystrophy: Clinical Assessment, Surgical Considerations and Recommendations for Treatment. J Clin Med. 2026;15(6):2116.
15. Elhamrawy A, Villalobos MA, Heydinger G, Corridore M, Tobias JD. Combined general and regional anesthesia for a patient with duchenne muscle dystrophy with an implanted left ventricular assisted device undergoing orthopedic surgery. J Med Cases. 2024;15(6):97.