Review Article

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Perioperative Anaesthetic Management in Emery-dreifuss Muscular Dystrophy: A Narrative Review

Anaesthesia Section

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ABSTRACT

Emery-Dreifuss Muscular Dystrophy (EDMD) is a very rare but also clinically important laminopathy that is accompanied by early contractures, weakness of the humeroperoneal muscles and progressive cardiac involvement such as conduction abnormalities, atrial arrhythmias, and dilated cardiomyopathy. These multisystem characteristics present special perioperative problems in EDMD, and thus, EDMD is a high-risk condition for anaesthesiologists. This is a narrative review summarising the existing evidence, also highlighting the overall picture of perioperative considerations in EDMD in the light of preoperative optimisation, intraoperative monitoring, anaesthetic pharmacology and postoperative care. Preoperative evaluation of EDMD requires meticulous cardiac assessment with the help of ECG, echocardiography, Holter monitoring, as well as pacemaker/Implantable Cardioverter-Defibrillator (ICD) interrogation, alongside respiratory evaluation, which proves beneficial for restrictive physiology. Intraoperatively, it is necessary to have rhythm surveillance, invasive haemodynamic surveillance, quantitative neuromuscular surveillance, and the presence of external pacing. Succinylcholine should not be used because of the risks of hyperkalemia and rhabdomyolysis, and the non depolarising agents also require careful titration. Total Intravenous Anaesthesia (TIVA) may be preferred over volatile anaesthetic agents in selected patients, and multimodal analgesia, which is safe. Despite the advantages of the regional and neuraxial practices like opioid sparing and cardiac stability, their implementation needs a thorough selection of patients and cardiac preparedness. In the postoperative period, the EDMD patients should be monitored at the ICU level to identify the arrhythmias, respiratory dysfunction, and delayed neuromuscular recovery. The present review highlights the need for adequately standardised protocols and future research to improve perioperative safety related to the management of EDMD patients.

Keywords: Arrhythmias, Cardiomyopathy, Neuromuscular blockade, Perioperative monitoring, Regional anaesthesia

INTRODUCTION

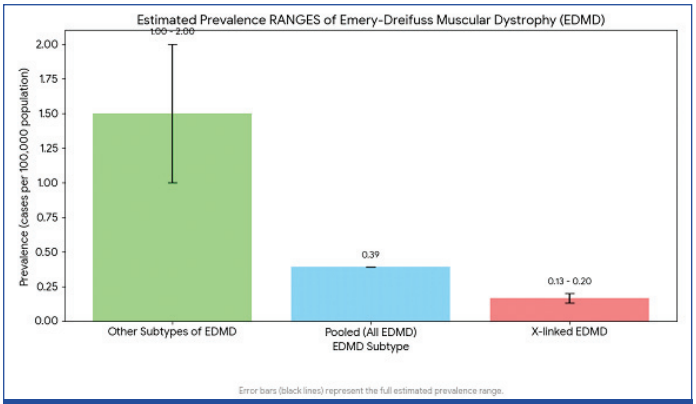
Emery-Dreifuss Muscular Dystrophy is a genetically heterogeneous neuromuscular disorder which is classically defined by the triad of early-onset joint contractures, a humero-peroneal pattern of slowly progressive muscle weakness as well as wasting, and prominent cardiac involvement, which includes conduction defects, arrhythmias, and dilated cardiomyopathy [1]. EDMD presents unique and clinically significant challenges for anaesthetic management due to this characteristic triad [1]. EDMD is very rare but clinically important because its cardiac complications can prove life-threatening, which may precede or outpace the skeletal muscle symptoms in severity [2]. The high prevalence of conduction abnormalities, atrial arrhythmias, and risk of sudden cardiac death makes perioperative cardiac optimisation crucial, while contractures and cervical spine rigidity may complicate airway management and positioning [2]. Additionally, EDMD patients may exhibit restrictive respiratory physiology and potentially altered responses to neuromuscular blockers and volatile agents [2]. EDMD is usually associated with nuclear envelope protein defects, out of which X-linked EDMD is most often caused by mutations in EMD (encoding emerin), also, autosomal dominant and recessive forms are mostly a result of mutations in LMNA (encoding A-type laminins) and, less frequently, other nuclear-envelope or associated genes [3]. These discoveries led to the placement of EDMD in a broader group of nuclear envelopathies or laminopathies [3].

The EDMD was recognised clinically as a distinct entity in the year 1960-1970, which was identified based on its distinct phenotype, early contractures of the elbows, Achilles tendons along with spine, humero-peroneal muscle wasting, and unusual cardiac pathology and was named after Emery AE and Dreifuss FE, who described familial cases and emphasised the cardiac risks [4]. The further era of EDMD began with the identification of mutations in EMD leading to X-linked EDMD and later recognition of mutations in LMNA that can also produce autosomal forms, other phenotype laminopathy [5]. EDMD is genetically as well as clinically heterogeneous, with a need for implications of family screening, cardiac surveillance, and consideration of device therapy (pacemaker/ICD) in affected patients [5].

The prevalence of EDMD estimated through meta-analysis of population-based studies, the pooled prevalence is 0.39 per 100,000 for all age groups [6]. X-linked form has a prevalence of approximately 0.13-0.20 per 100,000, but for other subtypes of EDMD, it is 1-2 per 100,000 [1]. A large study from the Indian subcontinent (involving clinical-genomic evaluation of 207 patients with inherited myopathies) identified one case of X-linked EDMD (EMD mutation) among the cohort [7]. In a more detailed study on nuclear envelopathy-related muscular dystrophies in an Indian cohort (16 patients), 4 patients were found with EMD (emerin) mutations and 11 with LMNA variants; out of all these patients, some had clinical features of EDMD [8]. This narrative review article aims to provide a comprehensive, evidence-based overview of various aspects of perioperative anaesthetic considerations as well as management strategies for patients with EDMD. Estimated prevalence ranges of EDMD Subtypes per 100,000 population are depicted in [Table/Fig-1].

Preoperative Evaluation and Optimisation in Emery-Dreifuss Muscular Dystrophy (EDMD)

The preoperative evaluation of EDMD implies a multisystem examination, but with a focus on cardiac and respiratory systems, since these two are the most significant factors of perioperative risks [1,9,10]. Given that EDMD is closely correlated with progressive atrial disease, conduction abnormalities, and dilated cardiomyopathy, a detailed cardiac work-up is necessary. ECG,



[Table/Fig-1]: Estimated prevalence ranges of Emery-Dreifuss Muscular Dystrophy (EDMD) subtypes per 100,000 population [1-6].

transthoracic echocardiography, Holter monitoring, and assessment of pacemaker/ICD functionality are necessary [11]. Although comprehensive cardiopulmonary assessment is recommended in patients having EDMD, the extent of preoperative investigations must be individualised according to clinical status as well as surgical urgency [10]. As EDMD is associated with progressive conduction system disease and atrial pathology, routine screening with electrocardiography and echocardiography is usually advised, while Holter monitoring or electrophysiological studies can be reserved for patients having symptoms, abnormal baseline ECG findings, history suggestive of arrhythmia, thereby improving the cost-effectiveness of evaluation strategies [10,11].

Current literature emphasises that optimisation must ideally occur during preoperative planning phase often several weeks before elective surgery allowing adequate time for cardiology consultation, adjustment of heart-failure therapy and device evaluation if required [12,13]. Prophylactic pacing is not indicated for all EDMD cases; but pacemaker implantation is recommended when clinically significant conduction abnormalities like sinus node dysfunction, Atrioventricular (AV) block, symptomatic bradyarrhythmias are identified given high prevalence of progressive conduction disease as well as risk of sudden cardiac death in patients [13,14].

In some cases where bradyarrhythmia or atrial standstill is suspected, perioperative availability of temporary pacing as well as defibrillation can be considered as a precaution even in patients without permanent device [13]. This tailored, multidisciplinary approach balances perioperative safety along with resource utilisation which remains important due to cardiac involvement into EDMD often progresses independently of severity of skeletal muscle disease [13,14]. Since EDMD patients might experience silent conduction blocks or atrial standstill, patients should be preoperative consulted with electrophysiology, early device interrogation, external pacing and defibrillation equipment should be available before anaesthesia [10,11].

The strategies of optimisation are aimed at stabilisation of cardiopulmonary functioning and prevention of the complications of the anaesthesia [10]. Adequate control of arrhythmia, anticoagulation management, along with heart-failure optimisation, which is done using beta-blockers, ACE inhibitors, or diuretics, when necessary, should be completed before starting the surgical operation [10]. Prophylactic pacemaker implantation in patients with conductive disease of more than 1.5 degrees, or atrial fibrillation, before elective surgery is also required to minimise the risk of perioperative arrest [10,15]. Respiratory evaluation is needed because of restrictive lung disease and reduced cough strength can also be present in EDMD; therefore, spirometry, sleep-disordered breathing assessment, and pulmonary physiotherapy are recommended when clinically necessary [16]. Nutritional evaluation, correction of electrolytes, and prevention of medications that lead to the development of rhabdomyolysis or malignant hyperthermia-like responses are also important preoperative aspects in EDMD [9,16].

Perioperative planning should focus on difficult airway positioning caused by rigidity in the cervical region, optimisation of mobility limitations which is posed by upper-limb contractures, and a personalised anaesthetic plan that minimises myocardial depression should be considered [10]. Regional techniques of anaesthesia should be used wherever possible, sedatives and neuromuscular blockers should be carefully titrated, and postoperative arrhythmias or respiratory depression should be closely monitored [17]. In total, evidence-based preoperative care, facilitated by cardiac optimisation, respiratory evaluation, and musculoskeletal factors, contributes dramatically to the decrease in perioperative morbidity in the EDMD patients [5,17]. Preoperative evaluation and optimisation in EDMD is explained in [Table/Fig-2] [1,5,9-11,15-17].

Domain	Key preoperative considerations	Rationale/ importance	References
Cardiac evaluation	• ECG, TTE, Holter monitoring • Assess pacemaker/ICD function • Electrophysiology consultation • Early device interrogation • Ensure availability of external pacing and defibrillation	EDMD is strongly associated with conduction abnormalities, atrial standstill, progressive atrial disease, and dilated cardiomyopathy; silent conduction blocks increase perioperative arrest risk.	[1,9-11]
Cardiac optimisation	• Control arrhythmias • Manage anticoagulation • Optimise HF with β-blockers, ACE inhibitors, diuretics • Consider prophylactic pacemaker if ≥1.5° conduction disease or AF	Stabilises haemodynamics, reduces risk of perioperative cardiac events and sudden bradyarrhythmic arrest.	[10,15]
Respiratory evaluation	• Screening for restrictive lung disease • Spirometry • Sleep-disordered breathing evaluation • Preoperative pulmonary physiotherapy	Weak respiratory muscles, reduced cough strength, and restrictive lung disease increase postoperative respiratory failure risk.	[16]
Respiratory optimisation	• Prehabilitation (breathing exercises) • Treat sleep apnoea if present • Ensure perioperative ventilatory support plan	Minimises postoperative hypoventilation and respiratory complications	[16]
Musculoskeletal and airway assessment	• Evaluate cervical rigidity for difficult airway • Assess upper-limb contractures affecting positioning • Plan optimal positioning strategies	Cervical rigidity may complicate intubation; contractures restrict safe positioning and increase injury risk	[10]
Medication review and metabolic optimisation	• Correct electrolytes • Nutritional assessment • Avoid drugs triggering rhabdomyolysis or MH-like reactions (e.g., depolarising NMBs)	Reduces risk of anaesthesia-related metabolic crises, rhabdomyolysis, and electrolyte-driven arrhythmias	[9,16]
Anaesthetic planning	• Favour regional anaesthesia when feasible • Carefully titrate sedatives and NMBs • Plan a myocardial-sparing anaesthetic strategy (e.g., TIVA)	Minimises myocardial depression, reduces arrhythmia burden, and prevents prolonged neuromuscular blockade.	[10,17]
Postoperative planning	• Continuous ECG monitoring for arrhythmias • Monitor respiratory function for hypoventilation • Prepare for early detection of conduction deterioration	High-risk of postoperative arrhythmias and respiratory compromise; early detection reduces morbidity	[17]

Overall importance	Evidence-based cardiac, respiratory, musculoskeletal, and metabolic optimisation significantly reduces perioperative morbidity in EDMD.	Multisystem stabilisation directly improves surgical outcomes.	[5,17]

[Table/Fig-2]: Preoperative evaluation and optimisation in Emery-Dreifuss Muscular Dystrophy (EDMD) [1,5,9-11,15-17].
ECG: Electrocardiography; TTE: Transthoracic echocardiography; ICD: Implantable cardioverter-defibrillator; AF: Atrial fibrillation; HF: Heart failure, ACE: Angiotensin-converting enzyme, NMBs: Neuromuscular blockers, MH: Malignant hyperthermia, TIVA: Total intravenous anaesthesia

Intraoperative Monitoring Strategies in Emery-Dreifuss Muscular Dystrophy (EDMD)

Intraoperative monitoring in EDMD requires to be monitored intensively since the affected patients are often associated with cases of conduction abnormalities, atrial arrhythmia and dilated cardiomyopathy, which often predispose to sudden haemodynamic collapse during anaesthesia [10,18]. It is also highly advised that continuous ECG monitoring with a focus on P-wave morphology, AV conduction intervals and arrhythmia detection be considered as the initial signs of high-grade AV block [19]. Intraoperative incidences of sudden bradyarrhythmias and asystole necessitate the use of external pacing pads, immediate availability of temporary pacing and constant interrogation of the device in patients possessing pacemakers or ICDs [12]. To monitor the invasive arterial blood pressure, it is recommended in patients with severe cardiomyopathy, arrhythmogenic load, or undergoing major surgery by providing beat-to-beat haemodynamic assessment when the rhythm becomes unstable [12,19].

In addition to standard monitoring, specific thresholds for intervention should be defined in patients having EDMD, usually due to conduction abnormalities; atrial standstill can progress abruptly during anaesthesia [20]. Continuous ECG monitoring must focus on detection of progressive PR interval prolongation (>200 ms), new AV block, and sustained bradycardia (<40-50 beats/min), which further prompt immediate evaluation as well as consideration of temporary pacing support in patients [21]. In patients having implanted pacemakers, implantable cardioverter-defibrillators, perioperative device interrogation and continuous rhythm surveillance are recommended; also, external pacing or defibrillation equipment must remain immediately available throughout the procedure [14,21].

The intensity of monitoring must also be tailored according to the complexity of surgery. For minor or short procedures, standard monitoring with continuous ECG, capnography, pulse oximetry and non invasive blood pressure measurement can be adequate in clinically stable patients without advanced type of cardiomyopathy [21,22]. However, for major surgery or procedures which are associated with significant fluid shifts or haemodynamic stress, invasive arterial blood pressure monitoring is recommended for allowing beat-to-beat blood pressure analysis as well as early detection of haemodynamic instability [21,22]. In selected high-risk patients who are having cardiomyopathy, unexplained intraoperative hypotension, intraoperative echocardiography (transthoracic or transesophageal) can provide valuable real-time information on ventricular filling, contractility and cardiac output [22,23].

Additionally, depth-of-anaesthesia monitoring using processed electroencephalographic techniques, inclusive of Bispectral Index (BIS) or entropy monitoring, can be useful to avoid excessive anaesthetic dosing, usually in patients having underlying cardiomyopathy or impaired physiological reserve [24]. Maintaining BIS values between 40 and 60 during general anaesthesia has been associated with adequate hypnosis while minimising anaesthetic-induced cardiovascular depression in the case of high-risk patients such as EDMD [24,25]. A multimodal monitoring strategy integrating rhythm surveillance, haemodynamic monitoring, respiratory as well as neuromuscular assessment and depth-of-anaesthesia monitoring thereby provides the safest perioperative approach for patients having EDMD [20,24].

Monitoring of the respiratory system and metabolism is also necessary since EDMD can be linked to respiratory muscle weakness, restrictive lung disease, and defective ventilatory reserve [26]. Continuous as well as rigorous monitoring of end-tidal CO_2 helps in the detection of early signs of hypoventilation, apnea, or insufficient neuromuscular recovery [26]. Core temperature monitoring, which helps to identify the hypermetabolic reactions, avoid stress caused by hypothermia and reduce postoperative respiratory compromise, must be utilised [26]. Quantitative neuromuscular monitoring is necessitated by changes in sensitivity to neuromuscular blockers and increased residual paralysis, as this has been reported in patients with laminopathies and other muscular dystrophies [23,26]. Full reversal of neuromuscular blockade must be given prior to extubation to reduce postoperative respiratory failure [26].

Intraoperative echocardiography (either Transthoracic (TTE) or Transesophageal (TEE) offers real-time assessment of ventricular activity, preload status and stroke volume as well as valvular pathology in patients with haemodynamic instability, preexisting cardiomyopathy, or otherwise unexplained intraoperative hypotension [27]. Echocardiography is especially useful in EDMD as atrial contraction loss or the development of atrial arrhythmias can significantly decrease the output of the cardiac muscle, and such alterations may occur abruptly during anaesthesia [27,28]. Altogether, a multimodal, high-resolution monitoring plan combining continuous rhythm analysis, invasive blood pressure surveillance, respiratory and neuromuscular monitoring, and selective perioperative echocardiography is a beneficial approach to preventing the morbidity and mortality of patients with EDMD [27].

Pharmacologic Considerations in Neuromuscular Blockade for EDMD

EDMD patients must be managed by keeping an eye on the same pharmacologic hazards described for other dystrophinopathies; depolarising neuromuscular blockers (succinylcholine) are generally avoided because they can provoke life-threatening hyperkalemia, rhabdomyolysis and myoglobin-related complications in patients with underlying myopathies [29,30]. Instead, when neuromuscular blockade is required, short-acting non depolarising agents and conservative doses can prove helpful [31]. Agents such as rocuronium (0.6 mg/kg i.v. for intubation with maintenance doses of 0.1-0.2 mg/kg), vecuronium (0.08-0.12 mg/kg i.v. for intubation with maintenance 0.01 mg/kg), atracurium (0.4-0.5 mg/kg i.v. for intubation with maintenance doses of 0.08-0.1 mg/kg), or cisatracurium (0.1-0.15 mg/kg i.v. for intubation) are commonly used intermediate-acting non depolarising neuromuscular blockers in anaesthesia practice [31]. Many patients show increased sensitivity and prolonged response, so quantitative usage of neuromuscular monitoring (TOF) along with careful objective assessment of recovery (TOF ratio ≥0.9) before extubation [31]. When residual blockade is suspected, pharmacologic reversal with neostigmine (0.04-0.07 mg/kg i.v. with an anticholinergic such as glycopyrrolate) or sugammadex (2-4 mg/kg depending on depth of blockade for aminosteroid agents like rocuronium or vecuronium) may be considered to ensure safe recovery of neuromuscular function [31,32].

The usage of TIVA with propofol and short-acting opioid/analgesic infusions (e.g. remifentanil) is an easier choice, which should be accompanied by opioid-sparing multimodal analgesia, prevention of excessive neuromuscular blockade, and perioperative ECG/arrhythmia monitoring (e.g. in the presence of EDMD cardiomyopathy/conduction disease) [33]. Preparation to treat hyperkalemia and rhabdomyolysis (when Individualised TIVA plans, judicious application of shorter-acting non depolarisers with neuromuscular monitoring, and aggressive perioperative cardiac surveillance on EDMD patients also proves beneficial [31,34]. Pharmacologic and anaesthetic considerations for patients with EDMD are mentioned in [Table/Fig-3] [29,30,31,33,34].

Aspect	Why/Rationale	Recommended practice	References
Avoidance of depolarising neuromuscular blockers (e.g., Succinylcholine)	Risk of life-threatening hyperkalemia, rhabdomyolysis, and myoglobin-related complications in EDMD and other dystrophinopathies	Completely avoid succinylcholine; choose alternatives	[29,30]
Use of non depolarising neuromuscular blockers	Patients often show increased sensitivity, prolonged response, and unpredictable recovery	Prefer short-acting non-depolarisers; use conservative doses; always use quantitative TOF monitoring; ensure TOF ratio ≥0.9 before extubation	[31]
Total Intravenous Anaesthesia (TIVA)	Safer profile for muscular dystrophies; avoids triggers associated with inhalational agents; provides stable control	Use propofol + short-acting opioid (remifentanil) infusions. Combine with opioid-sparing multimodal analgesia	[33]
Analgesia strategy	Opioid-related respiratory compromise and prolonged effect may be more pronounced in myopathies	Implement multimodal, opioid-sparing analgesia	[33]
Monitoring for cardiac complications	EDMD patients frequently have conduction abnormalities, arrhythmias, or cardiomyopathy	Continuous ECG and arrhythmia monitoring perioperatively	[33]
Risk of hyperkalemia and rhabdomyolysis	Underlying muscle membrane instability increases risk during anaesthesia.	Ensure availability and preparedness to treat hyperkalemia and rhabdomyolysis; avoid triggering agents	[29,30]
Overall anaesthetic plan	EDMD patients need individualised, cautious anaesthetic management due to neuromuscular and cardiac vulnerability	Use individualised TIVA plans, judicious short-acting non depolarisers with monitoring, and aggressive perioperative cardiac surveillance	[31,34]

[Table/Fig-3]: Pharmacologic and anaesthetic considerations for patients with Emery–Dreifuss Muscular Dystrophy (EDMD) [29,30,31,33,34].
EDMD: Emery-Dreifuss muscular dystrophy; TIVA: Total intravenous anaesthesia, TOF: Train-of-Four; ECG: Electrocardiogram; AV: Atrioventricular

Role, Advantages, and Limitations of Neuraxial and Regional Anaesthesia in EDMD

Neuraxial and regional techniques also offer clear perioperative benefits in patients with EDMD, as there is reduced systemic opioid requirement, preservation of spontaneous ventilation, along with provision of more stable haemodynamics compared with deep general anaesthesia, important in such a disorder where cardiac conduction issues and cardiomyopathy are common [26]. Several case reports of successful orthopaedic, general and obstetric surgeries have been done using the continuous epidural or spinal method in EDMD and have shown that neuraxial blocks are a feasible alternative when there is cardiac status is optimised and the availability of the necessary monitoring and pacing back-up [35]. In addition to neuraxial techniques, peripheral nerve blocks for upper and lower-extremity procedures (inclusive of femoral, sciatic, brachial plexus blocks) have also been reported as useful alternatives which allow surgery with minimal sedation while preserving spontaneous ventilation and avoiding need for neuromuscular blocking agents [26,36]. Concurrently, EDMD patients are often prone to conduction abnormalities or a cardiomyopathy and therefore any intervention that depends upon neuraxial sympathetic blockade needs to be weighed against the risk of bradycardia or hypotension and should not be attempted unless cardiology consultation has been obtained, with ECG being monitored and with continuous ECG monitoring and immediate pacing/defibrillation capability [30,35].

The safety and efficacy of regional techniques are susceptible to technical and pharmacologic limitations, which are particularly related to EDMD [37]. Neuraxial needles can be difficult to place and may have unpredictable spread of local anaesthetic (due to joint contractures, spinal deformities or previous spinal surgery), and clinicians should be ready to augment inadequate blocks or switch to general anaesthesia in cases where there is diminished local anaesthetic effect or altered duration in neuromuscular disorders [26,30]. Therefore, a structured risk-benefit assessment is usually recommended before selecting neuraxial or regional anaesthesia in the case of EDMD patients. Regional or neuraxial techniques can be preferred in cases when the surgical procedure allows a reliable block, the cardiac status of the patient has been optimised using appropriate monitoring or pacing backup available and the anaesthesia team is prepared well for rapid conversion to general anaesthesia if the block proves inadequate [26,30,38].

Moreover, although general anaesthesia is more complicated in EDMD because of avoidance of suxamethonium and because of adequate precautions in dosage/titration of non depolarising neuromuscular blockers, neuraxial anaesthesia also carries the risk of having conduction abnormalities which may occur during or after surgery, regardless of the method used [39]. Conversely, when regional blockade is technically difficult and cardiac instability is significant, a carefully planned general anaesthetic (often using TIVA along with avoidance of depolarising neuromuscular blockers) can be a safer alternative in case of EDMD patients [39]. The risk-benefit assessment thus favours neuraxial/regional approaches only when: (1) the procedure and anatomy of the patient make a reliable block; (2) the cardiac team is certain of the patient stability or is provides pacing; and (3) the anaesthesia team is ready for rapid conversion and provide invasive monitoring, a well-planned TIVA technique with airway and prepared pacing is preferable [33,39].

Postoperative Intensive Monitoring and Proactive Management for EDMD

Postoperative care for patients with EDMD must be proactive and tailored to their high cardiac and respiratory risk, these patients require monitored postoperative placement in an ICU setting with continuous ECG (with attention to P-wave and AV conduction changes) evaluation, ready external pacing capability, along with low threshold for telemetry-led escalation due to progressive conduction disease and malignant ventricular arrhythmias which are described in the early postoperative period [10,19]. Respiratory vigilance is also equally important; patients with reduced pulmonary reserve or chest wall contractures can be helped with early involvement of respiratory therapy, frequent bedside spirometry/ Peak Expiratory Flow Rate (PEFR), consideration of non invasive ventilatory support (CPAP/BiPAP) when indicated, and avoidance of prolonged residual sedative/opioid effects that can precipitate hypoventilation or atelectasis [40]. Analgesia must follow a proper multimodal, opioid-sparing strategy, which can thereby minimise respiratory depression while still controlling pain and facilitating early chest physiotherapy and mobilisation [36,41]. In cases when general anaesthesia is used, short-acting agents along with objective neuromuscular monitoring must be considered for guiding reversal and extubation [41]. Finally, proper clear handover documentation of baseline cardiac rhythm, device and pacemaker status, perioperative arrhythmias, as well as an individualised escalation plan (which includes who to contact, thresholds for pacing/ICD interrogation, also criteria for transfer to higher care), since early recognition and rapid intervention for bradyarrhythmia/heart block and respiratory compromise substantially reduce perioperative morbidity in EDMD [40,41]. Postoperative management strategies for patients with EDMD are highlighted in [Table/Fig-4] [10,19,36,40,41].

Future Directions for Anaesthetic Management of EDMD

Future directions in the anaesthetic management of EDMD must prioritise a shift from anecdotal evidence to standardised,

Domain	Recommendations	Rationale/Details	References
Cardiac monitoring	Intensive Care Unit (ICU) placement with continuous Electrocardiogram (ECG)	Monitor P-wave and AV conduction changes; early recognition of progressive conduction disease or malignant arrhythmias	[10,19]
	Ready external pacing capability	For rapid intervention in bradyarrhythmia/ heart block	[10,19]
	Low threshold for telemetry-led escalation	Promptly address perioperative arrhythmias	[10,19]
Respiratory care	Early involvement of respiratory therapy	Particularly important for patients with reduced pulmonary reserve or chest wall contractures	[40]
	Frequent bedside spirometry/PEFR	Assess lung function and ventilation status	[40]
	Non invasive ventilatory support (CPAP/BiPAP), if indicated.	Prevent hypoventilation or atelectasis	[40]
	Avoid prolonged residual sedative/opioid effects	Reduce risk of respiratory depression	[40]
Analgesia	Multimodal, opioid-sparing strategy	Minimise respiratory depression while ensuring adequate pain control	[36,41]
	Facilitate early chest physiotherapy and mobilisation	Supports pulmonary hygiene and recovery	[36,41]
Anaesthesia considerations	Use short-acting agents in general anaesthesia	Allows faster recovery and reduces residual respiratory depression	[41]
	Objective neuromuscular monitoring for reversal and extubation	Ensures safe extubation and optimal respiratory function	[41]
Handover and escalation	Clear documentation of baseline cardiac rhythm, device/ pacemaker status, and perioperative arrhythmias	Provides essential information for ongoing care	[40,41]
	Individualised escalation plan (contact points, pacing/ICD thresholds, criteria for higher care transfer)	Early recognition and rapid intervention reduce morbidity	[40,41]

[Table/Fig-4]: Postoperative management strategies for patients with Emery-Dreifuss Muscular Dystrophy (EDMD) [10,19,36,40,41].
EDMD: Emery-dreifuss muscular dystrophy; ECG: Electrocardiogram; AV: Atrioventricular; CPAP: Continuous positive airway pressure; BiPAP: Bilevel positive airway pressure; PEFR: Peak expiratory flow rate; ICD: Implantable cardioverter-defibrillator

evidence-based care, with a focus on three key areas: validation of preferred techniques, consensus guideline development, and large-scale data collection [36,41]. Anaesthetic choices currently favour TIVA and regional techniques to avoid the potential risk of rhabdomyolysis or malignant hyperthermia associated with volatile anaesthetics and succinylcholine [36]. Thus, future research must focus on validating the safety and efficacy of these preferred methods, particularly by investigating any reported local anaesthetic resistance [39]. Furthermore, given the rarity and profound cardiac risk associated with EDMD, the establishment of multicentre registries and large databases is essential, allowing the development of robust, consensus-driven, and multidisciplinary preoperative and perioperative guidelines that can specifically address the unique challenges of cardiomyopathy and conduction defects in patients [1,42].

Additionally, postoperative care requires careful planning as patients with EDMD can develop delayed respiratory compromise and cardiac conduction abnormalities [5]. The extended postoperative cardiac as well as respiratory monitoring is useful, usually after major surgery or general anaesthesia, with consideration of inpatient rather than ambulatory surgery in patients having significant cardiomyopathy or conduction defects [1,42]. Clear discharge criteria must include stable haemodynamics, absence of arrhythmias and adequate respiratory function, while selected low-risk patients undergoing minor procedures with regional techniques can be considered for outpatient management with appropriate follow-up [5,43].

Authors' contribution: AG: Contributed to the conceptualisation, literature review, data collection, and drafting of the manuscript; VC: Provided supervision, critically revised the manuscript for important intellectual content, and approved the final version; BS: Contributed to literature review, data interpretation, manuscript drafting, and formatting of tables and figures. All authors read and approved the final manuscript.

CONCLUSION(S)

The EDMD poses a considerable anaesthetic dilemma because it has the characteristic triplet of cardiac complications (conduction defects, arrhythmias), joint contractures and muscle weakness. Preoperative optimisation should be geared towards aggressive cardiac evaluation and stabilisation (e.g. prophylactic pacing). The intraoperative concern must focus on TIVA and preventing the use of succinylcholine, and attentive neuromuscular and continuous ECG monitoring of patient. In case of certain cardiac stability, regional anaesthesia can be used. The postoperative care requires a proactive, evidence-based and personalised ICU surveillance to identify cardiac and respiratory compromise at the earliest stages. Future efforts in research must establish a proper standardised consensus-driven guideline based on large-scale data and multicentre registries.

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