Int J Anaesth Crit Care. 2026;5(1):1–4.

International Journal of Anesthesia and Critical Care Case Report

Anesthesia in a Patient with Ehlers-Danlos Syndrome and Mast Cell Activation Syndrome—Case Report

Carlos Henrique Viana de Castro[1], Fábio Fantazzini Vieira[1,*], Felipe Cruz Jorge[1], Marco Túlio Froes[1], Marcelo Figueiredo[2]

[1]Department of Anesthesiology, Hospital Lifecenter de Belo Horizonte, Belo Horizonte, MG, Brazil

[2]Department of Plastic Surgery, Hospital Lifecenter de Belo Horizonte, Belo Horizonte, MG, Brazil

[*]Correspondence should be addressed to Fábio Fantazzini Vieira, fabiofantazzinivieira@gmail.com

Received date: November 16, 2025, **Accepted date:** January 23, 2026

Citation: Viana de Castro CH, Vieira FF, Jorge FC, Froes MT, Figueiredo M. Anesthesia in a Patient with Ehlers-Danlos Syndrome and Mast Cell Activation Syndrome—Case Report. Int J Anaesth Crit Care. 2026;5(1):1–4.

Abstract

The concurrent presentation of Hypermobile Ehlers-Danlos Syndrome (hEDS) and Mast Cell Activation Syndrome (MCAS) poses significant anesthetic challenges, balancing the risks of tissue fragility and autonomic dysfunction with potential anaphylaxis. We report the anesthetic management of a female patient with confirmed hEDS and MCAS undergoing mastopexy. Preoperative planning focused on immune stabilization and mechanical protection. Balanced general anesthesia was maintained with Sevofluorane and Propofol to avoid histamine-releasing agents. To minimize neuraxial risks associated with connective tissue laxity, multimodal analgesia was achieved via an ultrasound-guided intercostal nerve block. The procedure was completed successfully without anaphylactic, hemorrhagic, or positioning-related complications. This case highlights the importance of a trigger-free anesthetic technique, the utility of peripheral nerve blocks as an alternative to neuraxial anesthesia in hEDS, and the adaptability required in prophylactic protocols.

Keywords: Ehlers-Danlos syndrome, Mast cell activation syndrome, Anesthesia, Perioperative management

Introduction

Both Ehlers-Danlos Syndrome (EDS) and, more prominently, Mast Cell Activation Syndrome (MCAS) are systemic disorders with significant anesthetic implications. The aim of this case report is to present a clinical scenario in which both conditions coexist and to discuss the various perioperative considerations involved.

Case Report

We describe the case of a 36-year-old female patient scheduled for breast implant removal and mastopexy. Written informed consent was obtained from the patient for the publication of this case report. She had a known history of Ehlers-Danlos Syndrome hypermobile subtype and Mast Cell Activation Syndrome, under regular use of Ketotifen, Bilastine, Montelukast, Famotidine, Trazodone, Naltrexone, Cyclobenzaprine, and occasional Testosterone. Her surgical history included breast augmentation with prosthesis under intercostal block with sedation and liposuction under neuraxial block.

On preoperative evaluation: blood pressure was 110/60 mmHg, heart rate 98 bpm, BMI 19.4, and SpO_2 97%. Neck circumference measured 34 cm, with no signs suggestive of difficult ventilation or tracheal intubation; Naguib score was -0.91.

Laboratory tests showed hemoglobin 13.3 g/dl, platelet count 222,000/mm^3, fasting glucose 81 mg/dl, INR 1.0, activated partial thromboplastin time 27s (normal ≤32s), creatinine 0.67 mg/dl, and ECG with regular sinus rhythm without abnormalities.

The anesthetic plan prioritized drugs without histamine-releasing properties, as well as meticulous protection of skin and bony prominences. Prednisolone was administered 48h before surgery, and naltrexone was discontinued 3 days prior [1].

Medications and dosages for pre-induction, induction, and maintenance are detailed in **Table 1**. Due to the lack of intravenous H2 antagonists in our hospital, we relied on the patient's preoperative oral dose of Famotidine. Oral famotidine has a peak plasma concentration (Tmax) of 1–3 hours and an elimination half-life of 2.5–3.5 hours; therefore, administration on the morning of surgery provided sufficient therapeutic levels during the intraoperative period. Following induction, pulmonary ventilation was achieved via face mask, and endotracheal intubation was uneventful (Cormack-Lehane grade I).

The surgical procedure itself and the anesthesia occurred uneventfully, total time of 4 hours after the start of surgery. Neuromuscular block reversal was not required. After the performance of intercostal block, ultrasound scanning confirmed the absence of pneumothorax.

The patient was extubated in the operating room without complications. She was monitored in the Post-Anesthesia Care Unit (PACU) for 2 hours, where she remained hemodynamically stable with adequate pain control. She was discharged home on postoperative day 1 following an uneventful recovery. At the two-week follow-up, she reported no delayed hypersensitivity reactions or wound healing issues.

Discussion

The simultaneous presentation of Hypermobile EDS (hEDS) and Mast Cell Activation Syndrome (MCAS) poses a distinct challenge: the anesthesiologist must navigate the mechanical fragility of connective tissue while managing a hypersensitive immune response. While hEDS is often associated with autonomic dysfunction and difficult positioning, the superimposition of MCAS introduces the risk of unpredictable anaphylaxis. This case highlights the necessity of a "trigger-free" approach, prioritizing balanced anesthesia and multimodal analgesia to bypass common mast cell activators. According to the 2017 international consensus, EDS is classified into 13 subtypes, among which the classical (cEDS), hypermobile (hEDS), and vascular (vEDS) forms are most relevant [2]. These variants present multisystem involvement, particularly affecting skin, joints, vasculature, and visceral structures, directly influencing anesthetic management. Evidence in the literature is limited, mostly based on case reports and expert opinion, requiring individualized approaches guided by pathophysiological principles.

Anesthetic risk stratification must include identification of the EDS subtype (in this case: hypermobile), detailed history of bleeding, spontaneous hematomas, previous anesthetic failures, recurrent joint dislocations, and possible cardiovascular manifestations. Craniocervical instability should be evaluated with imaging in suspicious cases. Patients with vEDS require extreme caution due to the risk of spontaneous vascular rupture.

Although major vascular rupture is rare in hEDS, capillary fragility and easy bruising are common. Monitoring should preferably be non-invasive. Automated blood pressure devices

Table 1. Medications administered.

Drug	Dosage	Timing
Famotidine	20 mg PO	Morning of surgery (home)
Diphenhydramine	50 mg	Pre-induction
Cefazolin	2 g	Pre-induction
Dexamethasone	6 mg	Pre-induction
Magnesium sulfate	2 g	Pre-induction
Propofol	100 mg	Induction
Ketamine	25 mg	Induction
Sufentanil	25 mcg	Induction
Rocuronium	50 mg	Induction
Remifentanil	0.07–0.10 mcg/kg/min	Maintenance
Propofol	50 mcg/kg/min	Maintenance
Sevoflurane	1.2–1.5%	Maintenance
Metamizole (dipyrone)	2 g	Analgesia
Tramadol	100 mg	Analgesia
Ropivacaine 0.5%	200 mg	Intercostal block
Ondansetron	4 mg	PONV prophylaxis

should be used cautiously to prevent hematomas. Arterial cannulation and central venous access should be ultrasound-guided and performed by experienced clinicians. Positioning requires adequate padding to prevent pressure injuries, skin lacerations, and plexus injuries.

While Vascular EDS (vEDS) carries the highest mortality risk due to spontaneous arterial rupture, Hypermobile EDS (hEDS)—the subtype presented in this patient—poses distinct and often underappreciated anesthetic challenges. Unlike vEDS, where neuraxial and regional techniques are frequently contraindicated due to vascular fragility, hEDS patients generally tolerate these procedures, though with caveats.

Temporomandibular joint (TMJ) abnormalities and cervical instability increase the risk of difficult intubation. Fiberoptic bronchoscopy is recommended in high-risk cases. In extreme joint mobility the risk of TMJ dislocation during laryngoscopy is high and iatrogenic injury to the cervical spine or peripheral nerves during positioning may occur. Positive-pressure ventilation should be performed cautiously due to the risk of barotrauma and pneumothorax, particularly in vEDS.

General anesthesia may be performed with inhalational agents or total intravenous anesthesia. No specific pharmacokinetic alterations have been reported, but neuromuscular monitoring is recommended, especially in patients with muscle weakness. Succinylcholine should be avoided in immobilized patients. Regional anesthesia is contraindicated in vEDS due to the risk of epidural hematoma and dural rupture. In other subtypes, it may be considered on a case-by-case basis, preferably after preoperative spinal imaging.

Peripheral nerve blocks should be ultrasound-guided and performed cautiously due to hematoma risk. Cases of local anesthetic resistance have been reported, suggesting altered tissue diffusion. Regarding hemostasis, conventional tests (INR, aPTT) may be normal. Approximately 26% of patients exhibit subclinical platelet dysfunction [3]. DDAVP may be indicated in patients with bleeding history, and prophylactic tranexamic acid can be considered. Tourniquet use should be avoided as it may precipitate diffuse bleeding or compartment syndrome.

Postoperative monitoring should be extended, especially in vEDS. Main complications include delayed hematomas, dislocations, internal bleeding, and spontaneous organ rupture. Early mobilization may help reduce complications. Aggressive prevention of postoperative nausea and vomiting is crucial, as vomiting can precipitate esophageal rupture.

During the perioperative period, these patients are at increased risk of immediate hypersensitivity reactions due to exposure to multiple degranulation triggers. Triggers include psychological (anxiety, stress), pharmacological (neuromuscular blockers, antibiotics, opioids), mechanical (pressure, friction, tissue trauma), and environmental (temperature extremes). Release of preformed mediators—such as histamine, prostaglandins, leukotrienes, and proteases (e.g., tryptase)—can cause manifestations ranging from mild cutaneous symptoms (erythema, pruritus) to severe systemic reactions with cardiovascular collapse and anaphylaxis [4].

Perioperative strategy should focus on preventing mast cell degranulation by identifying and avoiding known triggers. Preoperative assessment must include a detailed history of prior anaphylaxis and identification of specific allergies.

Implementation of preventive measures has a significant impact on reducing complications. Scheduling surgery as the first case of the day can minimize anxiety; preoperative anxiolytics may prevent stress-induced degranulation. A calm environment and temperature-controlled operating room are essential. Strict intraoperative temperature control with active warming devices and warmed solutions helps prevent hypothermia as a trigger.

Anesthetic drug selection must avoid histamine-releasing agents. Benzylisoquinolinium neuromuscular blockers (Atracurium, Mivacurium) should be avoided, with Succinylcholine or steroidal agents (Rocuronium, Vecuronium, Pancuronium) preferred. Among intravenous agents, propofol, etomidate, and ketamine have favorable safety profiles. Synthetic opioids (Fentanyl, Remifentanil, Sufentanil) have lower mast cell–mediator release potential compared to morphine and meperidine [5]. Regional anesthesia does not increase adverse event risk in this population and may be a valid alternative when indicated.

Pharmacological prophylaxis is recommended. H1 and H2 antagonists should be administered pre-induction, with synergistic and safe profiles. Corticosteroids, despite lacking definitive recommendations regarding dose and timing, are frequently included in prophylactic regimens. Regular maintenance medications for mast cell stability should be continued until the day of surgery [5].

Intraoperative management should prioritize early recognition of mast cell degranulation signs and immediate treatment according to severity. Cardiovascular manifestations (hypotension, circulatory shock) are the predominant presentation and may occur even in the absence of cutaneous signs. Treatment should be titrated according to severity, with intravenous epinephrine reserved for severe reactions. Volume resuscitation with crystalloids or colloids is fundamental to counteract peripheral vasodilation and capillary leakage.

Conclusion

The concurrent presentation of Hypermobile Ehlers-Danlos Syndrome (hEDS) and Mast Cell Activation Syndrome (MCAS) creates a unique anesthetic challenge, requiring a strategy that balances chemical stability with mechanical protection. This case demonstrates that a successful outcome relies on three pillars: strict avoidance of histamine-releasing agents, rigorous prophylactic blockade of H1 and H2 receptors—adapting administration routes to institutional availability when necessary—and meticulous patient positioning to prevent iatrogenic joint injury. Furthermore, we highlight that while neuraxial techniques may pose risks in connective tissue disorders, ultrasound-guided peripheral nerve blocks can be safely employed as effective opioid-sparing alternatives. Ultimately, a multidisciplinary preoperative plan is essential to mitigate the dual risks of anaphylaxis and tissue trauma in this complex patient population.

Conflicts of Interest

The authors declare no conflicts of interest.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Acknowledgments

None.

References

1. Goel A, Kapoor B, Wu M, Iyayi M, Englesakis M, Kohan L, et al. Perioperative Naltrexone Management: A Scoping Review by the Perioperative Pain and Addiction Interdisciplinary Network. Anesthesiology. 2024 Aug 1;141(2):388–99.
2. Malfait F, Francomano C, Byers P, Belmont J, Berglund B, Black J, et al. The 2017 international classification of the Ehlers–Danlos syndromes. Am J Med Genet C Semin Med Genet. 2017 Mar;175(1):8–26.
3. Wiesmann T, Castori M, Malfait F, Wulf H. Recommendations for anesthesia and perioperative management in patients with Ehlers-Danlos syndrome(s). Orphanet J Rare Dis. 2014 Jul 23;9:109.
4. Lide B, Mcguire S, Liu H, Chandler C. Mast cell activation syndrome-anesthetic challenges in two different clinical scenarios. J Biomed Res. 2022 May 28;36(6):435–9.
5. Dewachter P, Castells MC, Hepner DL, Mouton-Faivre C. Perioperative management of patients with mastocytosis. Anesthesiology. 2014 Mar;120(3):753–9.