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Case Report

Perioperative Anesthesia Management in a Patient with Ehlers-Danlos Syndrome: A Case Report of Scleral Buckling Surgery

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ABSTRACT

This case report describes the anesthetic management of a 27-year-old man with hypermobile Ehlers-Danlos syndrome (hEDS) who underwent scleral buckling surgery for retinal detachment. The patient who had joint hypermobility and mild mitral valve prolapse required careful anesthetic management. The surgery, which was performed under general anesthesia with a laryngeal mask airway and EDS-tailored monitoring, was uneventfully accomplished in 85 minutes. The patient was discharged the next day, recovering well, emphasizing the importance of careful preoperative evaluation, correct positioning, and multidisciplinary care for patients with hEDS undergoing complex ophthalmic surgery.

Introduction

Ehlers-Danlos syndromes (EDSs) are a diverse group of inherited connective tissue disorders caused by mutations in genes that encode proteins involved in collagen synthesis, and they manifest with skin hyperextensibility, joint hypermobility, and tissue fragility [1–6]. The 2017 International EDS Classification describes thirteen unique subtypes of which one is hypermobile EDS (hEDS), the most common form, estimated to occur in around 1 per 5000 individuals [1].

Because of the higher incidence of airway problems, joint dislocation, postoperative bleeding complications and altered responses to anesthetics in patients with EDS [2], they represent particular challenges in perioperative management. Among EDS patients, ophthalmic surgeries should be carefully managed because of ocular fragility and increased retinal issues [7]. This case report presents a successful perioperative anaesthetic management of a hEDS patient for scleral buckling surgery (SBS) for RRD discussing the specific problems faced and their managements.

Case Report

Patient Information

The patient is a 27-year-old Caucasian man characterized by height 175 cm and weight 58 kg, with a BMI of 18.9 kg/m². He was diagnosed with hEDS at the age of 15 by clinical criteria and genetic testing was not available at his diagnosis.

The patient had developed acute retinal detachment because of which he had loss of vision and photopsia in the left eye. Symptoms were characterized by peripheral visual loss, twinkle-lights phenomenon, and gradual perimetric progression over a 48-h period before the examination. He has been diagnosed with hEDS at the age of 15, and he had undergone surgery for bilateral hip

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dislocations in infancy as also more than five episodes of bilateral shoulder dislocations that were treated conservatively with physiotherapy. He also had a correction of clubfoot at 3 years with satisfactory functional results. His medical history is further complicated by mild mitral valve prolapse with trace regurgitation, gastroesophageal reflux disease, and an anxiety disorder. The patient does not report a family history of connective tissue disease, genetic disorder or anesthetic issues. He does not have a family history of hypermobility or skin disorders, and his parents and siblings are normal. Clinically, he has trial of SSRIs for anxiety control for which he discontinued use because of gastrointestinal intolerance. He has also been under the care of a gastroenterologist and had two endoscopies which were negative. The patient lives independently and is a full-time worker with ergonomic accommodations for his joint issues. He did not use tobacco or illicit drugs and did not consume alcohol. He has no family support system and has not previously attended EDS support group meetings. The patient is a well-nourished, thin male. A prominent skin hyperextensibility, the ability to stretch his forearm more than 3 cm, together with mild skin fragility with atrophic scars are remarkable signs. His joint hypermobility is significant, with a Beighton score of 7/9. This has already been achieved in terms of dorsiflexing his 5th metacarpophalangeal (MCP) joints beyond 90° bilaterally, thumb-to-forearm movement on both sides, and hyperextension of the elbow joint to greater than 10° bilaterally and palm touching the floor with forward bending. Cardiovascular exam is significant for a midsystolic click, blood pressure 110/70 mmHg, heart rate 68 bpm. Respiratory examination shows clear breath sounds and normal effort. Airway assessment indicates a Mallampati II classification, adequate mouth opening, and mild temporomandibular joint hypermobility (Figure 1,2).

The diagnostic methods included an ophthalmological evaluation with a dilated fundus examination, optical coherence tomography, and B-scan ultrasonography. Preoperative assessments comprised a comprehensive medical history, physical examination, ECG, echocardiography, complete blood count, coagulation profile, and a basic metabolic panel. The anesthetic assessment consisted of airway evaluation, including Mallampati classification and neck flexion-extension measure- ment, and a cardiovascular assessment.

The issues which are particularly unique to EDS were the missing genetic verification (in particular in view of historical unavailability of a diagnostic test)—and instead diagnosis relied on clinical criteria, how one would assess bleeding risk when coagulation studies are normal/which may anyway hide potential functional abnormalities, and there was no validated preoperative evaluation tool specifically for patients with EDS.

The diagnosis of rhegmatogenous retinal detachment was established thanks to the characteristic clinical picture and imaging studies in a given patient. The patient was classified as having EDS hypermobility type (hEDS) if 2017 criteria hEDS, and the presence of joint hypermobility (Beighton scoring), absence of vascular/ classical features and skin involvement was included with the diagnosis. Alternative diagnoses considered and excluded were other connective tissue disorders like Marfan syndrome (excluded due to lack of aortic involvement and lens dislocation), benign joint hypermobility syndrome (excluded due to the presence of skin involvement and systemic features), and secondary causes of retinal detachment (excluded due to the typical appearance and lack of inflammatory signs). The patient has several favorable prognostic factors, including his young age, the hEDS subtype (which is less severe than vascular EDS), stable cardiovascular status, good baseline functional status, no severe bleeding history, and successful independent living.

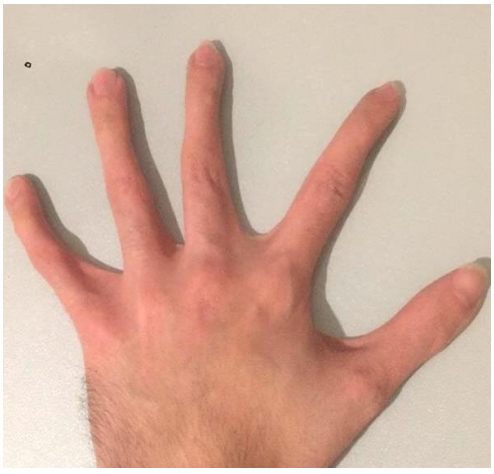

Figure 1- Hypermobile Hand in Ehlers-Danlos Syndrome

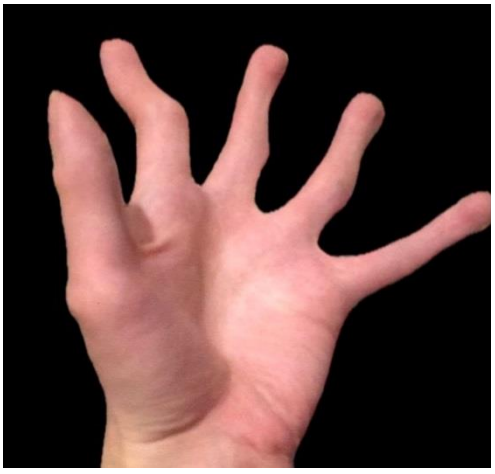

Figure 2- The hand with significant hyperextensibility and hypermobility (background removed for privacy)

However, there are also risk factors to consider, such as his joint hypermobility, which increases the risk of dislocation; skin fragility; potential resistance to local anesthetics; and a history of anxiety that requires careful psychological management.

The Panoramic Dental X-ray (Orthopantomogram - OPG) revealed abnormal root morphology, signs of premature moderate generalized periodontitis, and temporomandibular joint (TMJ) hypermobility. Electromyography (EMG) studies showed bilateral chronic mild L5 and S1 radiculopathy, with a history of axonal damage in the L5 distribution but no current active degeneration. Axonal injury to the deep peroneal nerves at both ankles is likely related to ankle instability and deformity, common features in EDS Radiographs demonstrated mild lumbar scoliosis, platyspondyly, loss of normal lordosis, segmental instability in the form of L4-L5 anterolisthesis and widened sacroiliac (SI) joints.

The patient does not have a history of anesthetic complications as he responds well to local anesthesia during dental procedures. As part of his past medical history, he had problems with wound healing, recurrent bilateral hip dislocation and multiple shoulder dislocations. He did not complain of arthralgia, chronic pain; he had no complaint of chronic fatigue. 3. He also experienced some dizziness, headaches, and GI symptoms that included occasional reflux despite 2 negative endoscopic examinations. While not showing signs of depression, he struggled with anxiety. According to the patient, his condition did not affect his daily functioning. Also; the patient had no notable history of inguinal hernia operation.

Written informed consent was obtained from the patient for the surgical procedure, including specific EDS-related risks and precautions, the anesthetic plan with a detailed discussion of airway management and positioning requirements, and the publication of this case report, including clinical details while protecting the patient's identity. The patient consented to academic publication and educational use of the case. All consents were documented in the medical chart, with both patient and witness signatures.

Due to difficulties in finding a suitable vein, intravenous access was established using a small-sized catheter. The interventions included general anesthesia with a laryngeal mask airway, neuromuscular blockade, and multimodal analgesia. Buckle surgery with cryotherapy for retinal tears was the surgical procedure. Preventive strategies consisted of specific positioning methods, soft tissue care protocols and postoperative nausea and vomiting (PONV) prophylaxis. Monitoring included cardiovascular monitoring, measurement of joint position, and bleeding surveillance. Premedication with midazolam(0.05 mg/kg),induction with propofol (1.5 mg/kg), fentanyl(2 mcg /kg) and atracurium (0.5 mg /kg) were used for intubation. Intra-operatively propofol (100–150 mcg/kg/min) was used for maintenance, along with an oxygen-air combination, and normal saline as a fluid regimen. Perioperative antiemesis was performed using ondansetron, and neostigmine and atropine were used for reversal agents. Analgesia was provided with acetaminophen and ketorolac. Changes to plan intraoperatively included slight increase of dose of propofol due to autonomic response, further padding repositioning for joint protection and avoidance ventilation pressure above 20 cmH2O to respect barotrauma.

Clinician-determined outcomes demonstrated an effective retinal reattachment under observation of ophthalmological examination, stable hemodynamics during the surgery and recovery period, good wound healing, and absence of joint dislocation or skin nothing. The patient complained minimal discomfort postoperatively (2-3/10 pain score on the Visual Analog Scale) and performed a quick return to baseline function. No perioperative complication including joint dislocation, skin injury or bleeding, airway injury, pneumothorax, hemodynamic instability or allergic reactions was observed.

The absence of local anesthetic resistance in his history, which was contrary to some literature reports, was an unanticipated finding. Additionally, despite identified risk factors, there was no postoperative nausea and vomiting.

Discussion

EDS is a group of genetic connective tissue disorders with defects in collagen synthesis, leading to symptoms like skin hyperextensibility, joint hypermobility, and tissue fragility. The overall incidence of EDS in the general population ranges from 1 in 10,000 to 1 in 25,000, with no ethnic predisposition [1, 6–8]. EDS has thirteen subtypes, with hypermobile EDS (hEDS) being the most common [4]. It is essential to consider the management of anesthesia in patients with EDS because many have joint hypermobility, and may sustain dislocations with positioning. Vascular EDS (vEDS) imposes other hazards due to tissue laxity and possibility of vascular involvement [1,5,9-10]. The main problems are local anesthetic resistance, neuraxial technique difficulties and respiratory complications [2-3,5,11–15]. Structural anomalies such as pectus deformities and scoliosis, complicating airway management and leading to reduced lung function, are also seen in other connective tissue disorders [16-17]. Good airway assessment, gentle patient placement and tissue handling (not stretching or tearing vessels), the ability to recognize immediate bleeding, continued monitoring for bleeding and close haemodynamic control are essential in order to render safe conditions and minimize complications [2,5,12–19]. This case contributes to the limited literature on

anesthetic management of ophthalmic surgery in EDS patients. Airway management, joint dislocations, and intraoperative bleeding complications have been previously described [3,12]. An LMA was successfully used, as previously advocated for the avoidance of airway injury in these patients [2, 12].

The successful outcome supports several key conclusions: (1) A comprehensive preoperative assessment is crucial for identifying and mitigating EDS-specific risks, (2) A laryngeal mask airway (LMA) can be safely used in hEDS patients when the airway assessment is favorable, (3) Meticulous positioning and tissue handling are essential and effective in preventing complications, (4) Standard anesthetic doses appear appropriate for hEDS patients without apparent pharmacokinetic differences, and (5) Multidisciplinary coordination enhances safety and outcomes. These conclusions are supported by the absence of complications despite the identified risk factors.

Conclusion

The main takeaway lessons from this case are both clinical and systemic. Clinically, the key lessons are that EDS patients can safely undergo complex procedures with appropriate preparation, risk mitigation through systematic assessment and planning is more effective than avoiding procedures, patient-specific factors are more important than diagnostic labels in determining management, and communication between specialties is crucial for optimal outcomes. Systemically, there is a need for standardized EDS anesthesia protocols, the value of genetic testing for diagnostic confirmation is highlighted, the importance of patient education and engagement in care planning is emphasized, and documenting successful approaches is beneficial for guiding future care.

The management approach's strengths lay in its thorough preoperative evaluation, collaborative multidisciplinary planning, and the implementation of evidence-based EDS-specific precautions, all of which contributed to a successful outcome without complications.

However, the approach has several limitations: it is based on a single case, which restricts its general applicability; the EDS diagnosis lacks genetic confirmation; there are no standardized EDS anesthesia protocols; long-term follow-up data is limited; and, very importantly, genetic testing was not available. It is also relevant to point out that EDS-related symptoms of the patient are rather mild, and they might therefore not reflect the challenges for clinicians dealing with more severely affected individuals.

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References

[1] 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;175(1):8-26.

[2] Laserna A, Nishtar M, Vidovich C, Borovcanin Z. Perioperative Management of Ehlers-Danlos Type III Syndrome Associated With Postural Orthostatic Tachycardia in Patients Undergoing General Anesthesia. Cureus. 2021; 13(11):e19311.

[3] Castori M. Surgical recommendations in Ehlers-Danlos syndrome(s) need patient classification: the example of Ehlers-Danlos syndrome hypermobility type (a.k.a. joint hypermobility syndrome). Dig Surg. 2012;29(6):453-5.

[4] Types of EDS - The Ehlers-Danlos Support UK [Internet]. [cited 2025 May 10]. Available from: https://www.ehlers-danlos.org/what-is-eds/information-on-eds/types-of-eds/

[5] Ehlers Danlos Anesthesia Considerations and Risks | The EDS Clinic [Internet]. [cited 2025 May 16]. Available from: https://www.eds.clinic/articles/anesthesia-risks-for-ehlers-danlos-syndrome-patients

[6] Ehlers Danlos syndrome - NYSORA [Internet]. [cited 2025 May 10]. Available from: https://www.nysora.com/anesthesia/ehlers-danlos-syndrome/

[7] Asanad S, Bayomi M, Brown D, Buzzard J, Lai E, Ling C, et al. Ehlers-Danlos syndromes and their manifestations in the visual system. Front Med (Lausanne). 2022; 9:996458.

[8] Ehlers-Danlos syndrome: MedlinePlus Genetics [Internet]. [cited 2025 May 10]. Available from: https://medlineplus.gov/genetics/condition/ehlers-danlos-syndrome/

[9] Im JS, Lim YH, Park JS, Lee SS, Kim KM. Rupture of abdominal aortic aneurysm after spine surgery in the patient with Ehlers-Danlos syndrome - A case report. Korean J Anesthesiol. 2010; 58(6):555-9.

[10] Lane D. Anaesthetic implications of vascular type Ehlers-Danlos syndrome. Anaesth Intensive Care. 2006; 34(4):501-5.

[11] Ohshita N, Kanazumi M, Tsuji K, Yoshida H, Morita S, Momota Y, et al. Anesthetic Management

of a Patient With Ehlers-Danlos Syndrome. Anesth Prog. 2016; 63(4):204-207.

[12] 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; 9:109.

[13] Bascom R, Dhingra R, Francomano CA. Respiratory manifestations in the Ehlers–Danlos syndromes. Am J Med Genet C Semin Med Genet. 2021;187(4):533–48.

[14] Schubart JR, Schaefer E, Janicki P, Adhikary SD, Schilling A, Hakim AJ, et al. Resistance to local anesthesia in people with the Ehlers-Danlos Syndromes presenting for dental surgery. J Dent Anesth Pain Med. 2019;19(5):261-270.

[15] Ohashi N, Furutani K, Ishii H, Baba H. [Perioperative brachial plexus injury caused by hyperabduction of the upper extremity in a patient with Ehlers-Danlos syndrome in the prone position]. Masui. 2012; 61(6):626–8.

[16] Athari M, Tajbakhsh A, Mirbolook A, Beheshtian M, Forghan M, Abtahi D. Management of scoliosis in a pediatric patient with lysyl hydroxylase-3 deficiency: a case report J Med Case Rep. 2024; 18(1):474.

[17] Glard Y, Launay F, Edgard-Rosa G, Collignon P, Jouve JL, Bollini G. Scoliotic curve patterns in patients with Marfan syndrome. J Child Orthop. 2008; 2(3):211-6.

[18] Kroese LF, Mommers EH, Robbens C, Bouvy ND, Lange JF, Berrevoet F. Complications and recurrence rates of patients with Ehlers-Danlos syndrome undergoing ventral hernioplasty: a case series. Hernia. 2018; 22(4):611-616.

[19] Wakabayashi R, Tanaka S, Tsuchiyama K, Yamamoto K, Maruyama Y, Numata K, et al. Anesthetic management of a patient with musculocontractural Ehlers-Danlos syndrome undergoing scoliosis surgery. JA Clin Rep. 2020; 6(1):46.