

Case Report

Anesthesia in a patient with Ehlers-Danlos syndrome: A case report

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Abstract

Ehlers-Danlos syndrome (EDS) is an inheritable group of connective tissue disorders characterized by joint hypermobility and instability, skin and vascular fragility and anomalies in various internal organs. Since collagen is found in multiple organ systems, distinct anesthesia considerations for each organ system have to be outlined as per patient symptoms. Anesthesia in EDS can be challenging due to involvement of different organ systems.

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1. Introduction

Ehlers-Danlos syndrome (EDS) is an inheritable group of connective tissue disorders characterized by joint hypermobility and instability, skin and vascular fragility and anomalies in various internal organs.¹ EDS primarily impairs collagen production and since collagen is found throughout the body in various organ systems, there are various types and degrees of symptoms. The incidence of EDS is 1:10000 to 1:25000 and it consists of multiple forms with different subtypes.¹ In 2017 an international classification system was established to categorize EDS in a systematic way.² Though each type of EDS has its own signs and symptoms, joint hypermobility and easy bruising are common in multiple types of EDS. In patients with EDS, anesthetic management requires a tailored approach.

2. Case Report

A 21-year-old female (165 cm, 74 kg) diagnosed to have EDS was scheduled for endoscopic septoplasty and adenoidectomy. She had complaints of nasal obstruction and difficulty in breathing. She was known to have thalassemia trait and diagnosed to have joint hypermobility syndrome- Ehlers-Danlos syndrome. She complained of pain in the right half of the body, swelling around the right shoulder, greater trochanter tenderness, myo-fascial and neck muscle tenderness. Her thumb in conjunction with flexion and extension of her wrist could touch her forearm. **(Figure 1)** Her skin was hyper-extensible, smooth and velvety. **(Figure 2)** Her

electrocardiogram was normal and 2 D echocardiography reported a left ventricular ejection fraction 55%, no regional wall abnormality, thickened mitral valve with trivial mitral regurgitation, trivial aortic regurgitation and mild tricuspid regurgitation. Pulmonary artery systolic pressure was estimated to be 30 mm of Hg. CT scan of paranasal sinuses reported mucosal thickening of maxillary and ethmoidal sinuses, pneumatization of crista galli and concha bullosa on both sides.



Figure 1: Thumb-to-forearm apposition demonstrating joint hypermobility



Figure 2: Demonstration of skin hyperextensibility in Ehlers-Danlos syndrome

In the operation theatre, routine monitors were attached and a blood pressure of 110/78 mmHg, heart rate 76 beats/minute and SpO₂ 100 were obtained. Anesthesia was induced with propofol, fentanyl and atracurium. Anesthesia was maintained with sevoflurane

in air oxygen mixture. Care was taken to avoid hyperextension of the neck during mask ventilation. Gentle laryngoscopy was done and excessive mouth opening was avoided. Intubation was done with 7.00 mm ID endotracheal tube. Patient was carefully positioned with adequate padding of pressure points. Hyperextension of shoulder and hip joints and cervical spine were avoided under muscle relaxation. Intraoperatively the hemodynamic parameters were generally stable. Excessive mouth opening was avoided for placement of retractor for adenoidectomy. At the end of surgery, temporomandibular joint was checked for any luxation. Extubation was uneventful and no adverse effects were reported during the postoperative period.

3. Discussion

Collagen production is primarily impaired in patients with EDS.³ Since collagen is found in multiple organ systems, distinct anesthesia considerations for each organ system have to be outlined as per patient symptoms. Therefore, preoperative history and physical examination are of utmost importance to formulate the anesthesia plan. The laboratory investigations are usually normal in these patients. Even though laboratory tests including INR, PTT and thromboelastogram are normal, 26% of these patients have platelet aggregation abnormality.⁴ Prophylactic tranexamic acid is recommended before surgery.⁵ Skin fragility has to be evaluated to plan placement of intra-venous lines and medical tapes. These patients are also prone to bruising due to repetitive inflation of non-invasive blood pressure cuff.¹ In patients with EDS subtype with vascular fragility placement of invasive arterial line can lead to vascular wall dissection. Adequate padding should be provided to prevent external tissue pressure. In EDS subtype with kyphoscoliosis and brittle cornea syndrome, care should be taken to prevent direct pressure on eyeball as it can lead to retinal detachment and even globe rupture.¹

In classical EDS cervical spine instability can lead to complications during intubation. Therefore, careful evaluation of cervical spine should be carried out prior to starting induction of anesthesia. Temporomandibular joint hypermobility increases the chances of subluxation and can make airway management difficult. Hence while opening the mouth, extra caution should be taken. Repeated intubation attempts can lead to mucosal injury and bleeding. Smaller sized tubes can decrease likelihood of mucosal injury. Patients with scoliosis may have restrictive lung disease and require lung-protective ventilation. Dysmotility of cricoarytenoid joint and lax vocal cords can lead to dysphonia.¹ In the postoperative

period, patients should be observed for pneumothorax, tracheal collapse and obstructive sleep apnea.

EDS patients are more prone to cardiac abnormalities like mitral valve prolapse and aortic root dilation. In patients having postural orthostatic tachycardia syndrome (POTS) adequate crystalloid infusion and early use of vasopressors may be required intraoperatively.⁶ There are increased chances of bowel perforation during endoscopic procedures. These patients are more prone to esophageal tear due to retching which can be minimized by adequate prophylaxis for postoperative nausea and vomiting.⁷ During laparoscopic surgeries, ventilation with higher inspiratory positive pressures can lead to pneumothorax.¹ Chest X-ray or sonography should be done postoperatively in these patients.

EDS patients are resistant to topical local anesthetics due to improper diffusion of local anesthetic because of presence of abnormal collagen.¹ Neuraxial anesthetic techniques may cause hematoma formation in patients with vascular EDS.⁸ Volatile and intravenous anesthetics and muscle relaxants are considered safe in these patients. Monitoring of neuromuscular blockade while emerging from anesthesia is recommended in patients with muscular weakness.¹ These patients are prone to chronic pain and complex regional pain syndrome and require multimodal pain management strategies. The risks and benefits of NSAID's should be considered before their use.

In our patient all these complications were considered. Gel mats were placed on the OT table to prevent undue pressure. Care was taken to prevent positional injuries and excessive mechanical stress. Excessive neck extension and mouth opening were avoided during intubation. Pressure control mode was used during ventilation to avoid excessive pressures during mechanical ventilation. Postoperatively nausea and vomiting were adequately controlled with ondansetron. Patient was comfortable postoperatively with a visual analogue scale reading of 1-3.

4. Conclusion

In conclusion, anesthesia in EDS can be challenging due to involvement of different organ systems. There is not enough literature to guide anesthetic management in these patients. Anesthesia in these patients has to be patient specific as per the organs involved. In addition, a database should be collected regarding perioperative management and complications in these patients for future guidance.

Author Contributions

Hemlata Kapoor: Manuscript writing and final approval of the manuscript.

Sachi Tripathi: Manuscript writing and final approval of the manuscript.

Declaration of Patient Consent

The authors certify that appropriate patient consent was obtained for publication of clinical information and images.

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Conflict of Interest

None.

Use of Artificial Intelligence (AI)

The authors declare that no artificial intelligence (AI) tools were used in the generation, analysis, interpretation, or writing of this manuscript.

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