Primary Anesthetic Challenges In Ehlers- Danlos Syndrome.

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Introduction

Ehlers-Danlos syndromes (EDS) are a group of related inherited disorders caused by primary genetic defects in the collagen supporting bones, skin and blood vessels characterized by hypermobility of joints and fragility of vascular wall and rarely cardiac valvular lesions. We describe primary anesthetic challenges of such a patient with EDS during general anesthesia (GA).

Case description

EDS are a group of inherited disorders of the connective tissue characterized by hypermobility of joints and skin fragility. In addition, patients may present with airway difficulty and congenital heart disease (CHD) with mitral valve prolapse and proximal aortic dilatation. A 15 year old physically active girl presented for arthroscopic resection of the right lateral retropatellar cyst with normal airway without CHD. GA was induced with propofol and oxygen/nitrous oxide with intermittent fentanyl. Perioperative course was uneventful.

FEATURES OF EDS

1. Hypermobility of joints
2. Delayed development of motor milestones
3. Soft elastic fragile skin
4. Bleeding in vascular EDS
5. Kyphoscoliosis and restrictive lung defect in kyphoscoliotic EDS
6. Hypermobility, hypotonia and hip dislocations in neonates and infants in Arthrochalasia EDS
7. Cardiac valvular abnormalities in cardiac-valvular EDS
8. Short stature and curved or bowed limbs in Spondyloepiphyseal dysplasia (SEED) EDS
9. Loose skin with sagging, wrinkles and redundant skin dermatosparaxis EDS
10. Thin cornea and other eye abnormalities in brittle cornea syndrome EDS
11. Hypotonia and contractures in musculocontractural and myopathic Ehlers-Danlos syndrome

Cardiac Anomalies in EDS

1. Mitral valve prolapse
2. Mitral regurgitation
3. Tricuspid regurgitation
4. Aortic root dilation
5. Aortic regurgitation
6. Cardiac conduction abnormalities

Cardiac involvement is uncommon in EDS

References:
1. Ehlers-Danlos syndrome. Genetics Home Reference
2. The Ehlers–Danlos Society

Conclusion

The primary anesthetic concern during GA is airway management to avoid excessive neck movement. We achieved this goal with the placement of an LMA.