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"Primary Anesthetic Challenges In Ehler- 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 retropatellar cyst with normal airway without CHD. GA was induced with propofol and anesthesia was maintained with a laryngeal mask airway (LMA) with sevoflurane /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 kyphoscoloitic EDS
6. Hypermobility, hypotonia and hip dislocations in neonates and infants in Anthrochalasia EDS
7. Cardiac valvular abnormalities in cardiac-valvular EDS
8. Short stature and curved or bowed limbs in Spondylodysplastic 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

CASE DISCUSSION

Classification of EDS as per 2017 International diagnostic criteria [2] and mode of inheritance

- Classical (AD)
- Classical like (AR)
- Cardiac-valvular type (AR)
- Vascular (AD)
- Hypermobile (AD)
- Arthrochalasia (AD)
- Dermatosparaxis (AR)
- Kyphoscoliotic (AR)
- Brittle Cornea Syndrome (AR)
- Spondylodysplastic (AR)
- Musculocontractural (AR)
- Myopathic EDS (AD/AR)
- Periodontal EDS(AD)

AD=Autosomal dominant, AR= Autosomal recessive

Cardiac Anomalies in EDS [3, 4]

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

Anesthetic challenges in EDS [5]

1. Hypermobility of spine may result in cervical spine instability
2. Avoid excessive cervical movement is the primary goal during airway management
3. No guidelines exist with regard to GA and airway management
4. No guidelines exist on neuraxial blockade.
5. Neuraxial blockade may result in hematoma in vascular type of EDS

Airway management technique to minimize cervical movement in EDS

1. Use laryngeal mask airway(LMA)
2. Video laryngoscopy
3. Fiberoptic intubation
4. Cervical spine immobilization during direct laryngoscopy

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.

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