Anaesthetic Management of a Patient Having Ellis-Van Creveld Syndrome

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Abstract: Ellis-Van Creveld Syndrome is rare Aotossomal recessive condition characterized by short limb dwarfism, polydactyly, abnormal development of finger nails and congenital cardiac defects in 50% of the patients. The orofacial manifestation includes multiple gingivolabial fraenula, dental anomalies, hypodontia and malocclusion. The syndrome can be caused by mutation in EVC gene or EVC2 gene present in chromosome 4p16.

Keywords: Ellis-Van-Crevald syndrome, polydactyly, Chondroectodermal dysplasia

1.Case Report

A 3 yr old female, weighing 11 kg came with complains of dental caries for 5 months and was posted for pulpectomy. She was the first child of nonconsanguineous and normally developed parents. Her birth was full term normal vaginal delivery.

She had polydactyly in all 4 limbs, short limbs, conical teeth and multiple missing teeth.

On examination loud S1 and systolic murmur was heard in mitral region.

Her past medical history revealed that she was operated for AV canal repair and coronary sinus unroofing with left SVC ligation for severe mitral regurgitation at the age of 6 months.

She was a known case of Ellis-Van Creveld Syndrome and was posted for pulpectomy.





2. Preopertively

Her vitals and labs were normal except for 2D ECHO which showed mild PAH, severe AR, grade 1 right AV Regurgitation and moderately dilated LA. Her airway assessment was normal.

She was on T. Enalapril 2.5 mg BD and T. Furosemide 2.5 mg BD.

Glycopyrrolate 0.004 mg/k, Midazolam 0.05mg/kg and Inj. Fentanyl 2 mcg/kg was given as premedication.

3.Intraopratively

She was induced with Inj Propofol 2mg/kg and Inj Atracurium 0.5 mg/kg and intubated with north pole uncuffed ET 4.5 and maintained on oxygen, Air, Sevoflurane and Atracurium. Inj Neostigmine 0.05mg/kg and Glycopyrrolate 0.008mg/kg was given and she was extubated.

Postoperatively she was sent to ward and discharged after one day.



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4.Discussion

The primary goal of anaesthetic management is thorough evaluation of airway, skeletal deformity and cardiac anomaly.

The aim of intraoperative management is control of airway, prevention of aspiration, fluctuation in BP, barotrauma, myocardial depression, worsening pulmonary hypertension and heart failure. Ventilatory management is directed towards stable airway pressure, normoxia and normocapnia.

Post operative management should constitute adequate analgesia and prevention of adverse cardio respiratory events.

References

- Guha A, Malhotra R, Saxena R, Keshri VK. Ellis van creveld syndrome: Cardiac anomalies and anesthetic implications. Ann Card Anaesth. 2023 Jul-Sep;26(3):346-348. doi: 10.4103/aca.aca_166_22. PMID: 37470539; PMCID: PMC10451126.
- [2] Fernández-Meré LA, Alvarez-Blanco M, Jorge-García J, Martínez-Suárez MA. Tratamiento anestésico de una paciente con síndrome de Ellis-Van Creveld [Anesthesia in a patient with Ellis-van Creveld syndrome]. Rev Esp Anestesiol Reanim. 2010 Oct;57(8):528-31. Spanish. PMID: 21033459.
- [3] Abeles AI, Tobias JD. Anesthetic implications of Ellis-van Creveld syndrome. J Clin Anesth. 2008 Dec;20(8):618-21. doi: 10.1016/j.jclinane.2008.06.012. PMID: 19100937