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Cord Untethering in a Patient with VACTERL Syndrome

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Introduction

The incidence of VACTERL syndrome varies between 1; 10,000 and 1:40,000.¹ 90% of the cases are sporadic and 10% patients have a first degree relative with this association.² 40-80% of the VACTERL infants have congenital heart disease.² Association between VACTERL and Klippel-Feil (KF) syndrome. has not been described.³ The KF is slightly more common in females than males. The incidence of KF worldwide varies between 1:40,000-1:42,000.³ Mutations in the GDF6, GDF3 or MEOX1 gene may result in Klippel-Feil syndrome. These genes are involved in bone development.³

Case Discussion

A 7-year-old female with VACTERL syndrome, in the setting of global developmental delay, s/p repair ASD, VSD and PDA in infancy, and asthma underwent cord untethering for thickened philum terminale and spina bifida occulta.

Her presenting symptoms that necessitated surgery were lower extremity pain and urinary incontinence. Extra precaution was taken during airway management in the setting of Klippel-Feil (KF) syndrome. She was intubated with direct laryngoscopy. A remifentanil and propofol infusion was utilized to facilitate neurophysiological monitoring and the perioperative course was uneventful.

Characteristics of VACTERL Syndrome

Type	Incidence	Examples
Vertebral	17%	Scoliosis, vertebral defects
Anal	12%	Imperforate anus malrotation, duodenal atresia
Cardiac	20%	VSD, PDA, tetralogy of Fallot, ASD, right-sided aortic arch
TE Fistula		Tracheoesophageal fistula: Types: A-F (C most common)
Renal	16%	Renal agenesis/dysplasia, hypospadias, polycystic / horseshoe kidney
Limb	10%	Radial anomalies, polydactyly, lower-limb defects

Characteristics of Klippel-Feil Syndrome

Klippel Feil ¹	Symptoms	Percentage
TRIAD	Kyphosis / scoliosis	60
Short neck	Renal disease	35
	Synkinesis or movement	20
Low neck hairline	Sprengel deformity + torticollis	30
	Congenital heart disease	4 - 14
Limited Neck ROM	Brainstem lesion	<10
	Congenital cervical stenosis	<10

Effects of Anesthetics on evoked potentials
VEP > MEP > SSEP > BAEP

Anesthetic agents	Amplitude	Latency
IV and inhalational agents	Decreases	Decreases
Etomidate	Increases	Increases
Ketamine	Increases	No change
Nitrous Oxide	Decrease	No change

Management of VACTERL

- 1- Airway management plan and backup
- 2- Aspiration prophylaxis + gastric decompression
- 3- Review of ECHO for cardiac anomalies
- 4- Maintain spontaneous ventilation
- 5- Induce with ketamine and atropine (salivation+vagal tone) vs. sevo inhalation induction vs. awake intubation
- 6- Avoid positive pressure ventilation (aspiration)
- 7- Post extubation croup and OSA

References

- [1] Davis L. et al. Carpal tunnel syndrome in children. *Pediatr Neurol.* 2014 Jan;50(1):57-59.
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 [3] Mukund R. Thatte et al. Compressive neuropathy in the upper limb. *Indian J Plast Surg.* 2011 May-Aug; 44(2): 283-297.