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# Difficult Airway Management in a Patient with Klippel- Feil Syndrome

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## INTRODUCTION

Klippel-Feil syndrome is primarily characterized by a severely limited cervical range of motion as a direct result of multi-level cervical and in some cases thoracic vertebral fusion with anticipated difficult intubation.

## CASE DESCRIPTION

A 32-year-old Indian male with a history of Klippel -Feil type II who presented for anterior and posterior multilevel cervical fusion because of a fall. The patient suffered a chance fracture of C6-C7 and was placed in a cervical-collar for cervical stability. There were no neurological deficits.

Preoperatively the patient was hemodynamically stable and denied any other past medical history. However, he was unable to open his mouth and stated that he had been unable to open his mouth since he was a young child. He ate by placing soft food behind his right molars from the side of his mouth but did admit this was not always easy evident by his weight of 33Kg. Pertinent on physical exam was a kyphotic cervical curve with a slight rightward deviation and a cervical collar in place.

Preoperatively the patient was given glycopyrolate and oxymetazoline was applied to both nares. Next, dexmedetomidine was started with an initial bolus of 1mcg/kg followed by an infusion of 0.7mcg/kg/hr. An awake nasal fiberoptic intubation with nasal RAE tube after adequate topicalization with lidocaine with patient breathing spontaneously. After successful management of airway anesthesia was maintained with total intravenous anesthesia with infusions of propofol, remifentanil and dexmedetomidine, in addition to intermittent boluses of fentanyl.

Patient underwent anterior C3-T2 and posterior C3-T3 fusion with somatosensory and motor evoked potential monitoring throughout the surgical procedure.



## DISCUSSION

Klippel-Feil syndrome is characterized by the fusion of two or more cervical vertebrae. The mode of inheritance can be autosomal dominant or recessive. Females are affected more than males. Incidence is 1:40,000 <sup>1</sup>

Autosomal dominant: Caused by mutations in the GDF6 or GDF3 genes. Mutations in one copy of the gene is sufficient to cause the disorder.<sup>1,2</sup>

Autosomal recessive: Caused by mutations in the MEOX1 gene. Mutations in both copies of the gene are necessary to cause the disease.<sup>1,2</sup>

### Characteristic features of Klippel- Feil syndrome <sup>2</sup>

- Short, possibly webbed neck
- A low posterior hairline
- Limited mobility of the cervical spine

### Classification of severity <sup>2</sup>

- Type 1 : Patients with fusion of C2 and C3 with occipitalization of the atlas
- Type 2: Patients with long fusion below C2 with an abnormal occipital-cervical junction
- Type 3: Patients with presence of thoracic and lumbar abnormalities in addition to type 1 and 2

### Anesthetic considerations <sup>2</sup>

1. An unstable cervical spine may lead to neurological insult with head and neck manipulation during intubation
2. Possible difficult mask ventilation
3. As cervical fusion is progressive, a prior successful intubation with direct laryngoscopy may not assure easy repeat intubation with direct laryngoscopy
4. Use of awake fiberoptic with dexmedetomidine and ketamine is preferred

### Associated Anomalies<sup>2</sup>

- Atlantooccipital fusion, Skull deformities, Facial asymmetry
- Hearing loss, Cleft lip, Micrognathia, Oligodontia
- Torticollis, Laryngeal cartilage malformation with voice abnormalities
- Scoliosis, Genitourinary abnormalities
- Sprengel's deformity (scapulae ride high on the back)
- Synkinesia, Sacral agenesis
- Cardiovascular abnormalities (VSD)

### REFERENCES

1. Klippel Feil Syndrome, Genetics Home Reference
2. Almeida-Chen GM, Chapter 72, Klippel – Feil Syndrome, Hand book of pediatric anesthesia, Lange publications.