Klippel Feil Syndrome: A challenge to the Anaesthesiologists

**Abstract:** Klippel-Feil syndrome (KFS) is a rare syndrome characterized by a classic triad comprising a short neck, a low posterior hairline, and limited neck mobility due to fused cervical vertebrae. We hereby report anesthetic management for pyeloplasty of KFS patient with micrognathia and restricted mouth opening. A careful preoperative evaluation to exclude other anomalies associated with KFS was done. The patient had restricted mouth opening, retrognathia, and limited cervical mobility due to cervical vertebral fusion, with Sprengel deformity, aortic arch abnormality, sensory neuronal hearing loss and urologically pelviuretric obstruction on both sides. As difficult intubation was predicted, video laryngoscopy was our first choice for gaining control of the patient’s airway. In the operating room, tracheotomy equipment was kept ready if a perioperative surgical airway control was required. Surgery was uneventful. Careful preoperative examination and preparation for difficult airway management are important for KFS patients with micrognathia.

**Keywords:** Klippel Feil Syndrome, challenge to the Anaesthesiologists.

**INTRODUCTION**

Klippel-Feil syndrome (KFS) was first reported by Maurice Klippel and Andre Feil in 1912 in patient with congenital fusion of cervical vertebrae (Jones, K.L. 1997). In KFS, classically there is a triad of short neck, a low posterior hairline restricted motion of the neck due to fused cervical vertebrae, especially of lateral bending. All the three elements are present in fewer than 50% of cases. KFS occurs in one of every 42,000 births, of which females comprise of 60% of cases (Dikova, M. et al., 1996). It is associated with Sprengel deformity, high scapula, scoliosis, congenital heart disease, urinary tract anomalies and hearing loss in 30% of cases (Paul, W. et al., 2003). The anatomical characteristics of KFS have significant importance for airway management because it has a potential for difficult intubation (Khawaja, O.M. et al., 2009).

**CASE REPORT**

A 17 year old male attended outpatient department of surgery of MGIMS on 6th August 2020 with torticollis of left side. His height was 160 cm, weight 44 kg was scheduled for pyeloplasty for left pelviuretric junction obstruction. He was admitted with easy fatigability and abdominal discomfort on off since 1 week, which was dull aching type at right lumbar region. There was no significant antenatal / birth or family history of any congenital anomaly.
On examination- He had a small mandible and retrognathia, with an overjet (labial surface of the most prominent incisor to the labial surface of the mandibular incisor) of 2 mm. His cervical extension was reduced. Airway examination revealed that his Mallampati score was class 3. Mouth opening was two and half fingers, his temporomandibular joint acted only as a hinge, and it was not possible for it to glide forward. Spirometry showed slight restrictive ventilatory impairment resulting from thoracic hypoplasia and FEV1 of 73%. His vital signs, electrocardiogram, and blood tests were within normal limits.

Other findings included hypoplasia and asymmetry of the clavicle and scapula, with limited ability to raise his arms. Orthopedic examination revealed severe limitation of cervical mobility, with only 30 degrees of head flexion and extension and 30 degree of lateral rotation possible, due to fusion of the C4–5 vertebrae. However, the normal range of neck motion required for endotracheal intubation during anesthetic induction was unlikely to cause any neurological damage, as her X-ray showed no basilar invagination or cervical cord compression.

Neurosurgical examination demonstrated no atlantoaxial subluxation, no spinal compression or cervical unsteadiness. His magnetic resonance imaging and echoencephalograph revealed no cerebral abnormalities, and he had no neurological symptoms. He did not complain of either spontaneous pain or pain on motion in the back of the head and neck. His CT thorax showed bovine aortic arch and aberrant origin of left subclavian artery or arteria lusoria.
His 2D echo showed no LVH, no RWMA, valves normal, LVEF-65%, no pulmonary arterial hypertension, no ASD, PDA or coarctation.

Urological examination revealed left sided dilatation of renal pelvis with abrupt narrowing of pelviureteric junction possibly due to left sided pelviureteric junction obstruction.

Otolaryngological examination showed that microtia and retracted tympanic membrane. Pure tone audiometry showed bilateral mixed severe hearing loss and hearing aid trial was advised. Indirect laryngoscopy showed that bilateral vocal cords were mobile and there was no pooling of saliva.

ANAESTHESIA PLAN
We considered a multiple-contingency plan for induction of anesthesia and airway management. In view of the micrognathia and cervical spine deformity, we planned a video laryngoscopy. Written informed consent for the procedure was obtained from the patient and his parents, after explaining the risks involved in securing his airway. Before surgery, an intensive care unit bed was reserved for him in the eventuality of respiratory failure due to upper airway obstruction in the postoperative period. As a precaution, tracheotomy equipment was always kept ready in the operation room.

Patient was then taken inside operation theatre and multipara monitors were attached, 18 gauge IV cannula was secured and IV fluid ringer lactate was started. Premedications of inj glycopyrrolate 0.2mg iv and inj midazolam 1mg iv was given. Patient was preoxygenated with 100% oxygen for 5 minutes. Inj fentanyl 100mcg was given and induction was started with inj propofol 100mg was given slowly. Inj succinylcholine 100 mg iv given. Videolaryngoscopy was performed by an experienced anaesthesiologist. Airway assessment revealed that Cormack and Lehane grade was 2b, defined as only arytenoid cartilages were visible. Anaesthesia was maintained with air, oxygen, and sevoflurane (fraction of inspired oxygen [FiO2] ½ 0.4) under controlled ventilation. Left Anderson Hynes Desmembered pyeloplasty was done with right lateral poisoning given with adequate padding of the bony prominences. The surgery lasted for 1 hour 45 minutes, at the end of which the patient was extubated with no difficulty when he was completely awake. After extubation, he did not suffer respiratory failure and had an uneventful postoperative course.

DISCUSSION
KFS patients are of 3 types: type 1, cervical vertebrae with extensive fusions at many levels; type 2, those with fusions at only 1 or 2 cervical intervertebral spaces; and type 3, those with fusions in the cervical spine accompanied by fusion in the lower lumbar spine. Patients with type 2, which is the most common form of KFS, including our patient, most often have anatosomal dominant inheritance pattern. (Samartzis, D. et al., 2006; & Tracy, M.R. et al., 2004) KFS can be associated with a number of other organ system anomalies, including renal dysfunction (64%), scoliosis (60%), deafness (30%), Sprengel’s scapula deformity (25–35%), congenital heart disease (4.2–14%), mental deficiency, pulmonary disability, and cleft lip and palate (Bhandari, S., & Farr, M.J. 1996; Hase, Y. et al., 2014; Jones, K.L. 2006; Widgerow, A.D. 1990; Naguib, M. et al., 1986; & Hatano, K. et al., 1979). KFS is a malformation sequence that start at an early weeks of gestation usually the fourth or beginning of the fifth week of fetal life (Duncan, P.A. 1977). KFS encompasses an increasing constellation of clinical features associated with abnormalities of other organ systems may be present (Duncan, P.A. 1977; & Widgerow, A.D. 1990). KFS appears to be failure due to abnormal fusion of the cervical vertebrae and palatal shelf, causing cleft palate or retrognathia, and associated anomalies around the oral area (Bhandari, S., & Farr, M.J. 1996). Patients with KFS are at more risk of development of cervical instability, increased risk of neurologic damage with minor trauma or during laryngoscopy, intubation, and positioning (Naguib, M. et al., 1986). Obstructive sleep apnea can develop due to mandibular hypoplasia with KFS due to upper airway obstruction (Dove, N. et al., 2006). Teoh, D.C.A., & Williams, D.L. (2007) described associated craniofacial anomalies, such as mandibular malformations and micrognathia, may make oxygenation and ventilation difficult in these patients. Hence, understanding of anatomical and pathological changes associated with these congenital anomalies is very important for successful anesthetic management of these patients.

Patients with KFS pose many problems, one of which is difficult airway. In a previous case report of surgery in a KFS patient, the patient required emergency tracheostomy postoperatively for complete upper airway obstruction, although a blind oral intubation was successful after several attempts (Shudou, Y. et al., 2007). In another case, many attempts at endotracheal intubation were associated with complications such as hypoxia, laryngospasm, and bradycardia, which required cardiac massage, and even abandonment of the surgical procedure because of unsuccessful endotracheal intubation (Skakai, H. et al., 2001). Our KFS patient had micrognathia in addition to limitation of neck mobility due to cervical vertebral fusion. Although his mouth opening was about 30 mm with only a hinge action of mandibular movement at the temporomandibular joint. Video laryngoscopy was our first choice for securing the airway. As we predicted difficult intubation, we prepared for tracheostomy, and also, we had the other equipment for alternative airway management such as a laryngeal mask and percutaneous tracheal puncture needle (Papagiorgakis, M.J. et al., 2003). In conclusion, careful preoperative examination...
and preparation for difficult airway management are important for KFS patients with micrognathia.

REFERENCES


