



## Case Report

# Anaesthetic considerations in paediatric klippel feil syndrome- A case report

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### ABSTRACT

Klippel feil syndrome is a rare skeletal disorder primarily characterized by abnormal union or fusion of two or more cervical vertebrae which is often associated with cervical canal stenosis. This results in a short neck, a low hairline at the back of the head, a limited range of motion in the neck and infrequently an atlanto-axial instability.

Here we report a case of type 3 paediatric Klippel Feil syndrome who underwent Sprengel deformity repair in prone position and its anesthetic implications.

A careful assessment of airway and an algorithmic approach towards management is needed in children with KFS. The various spine deformities associated with KFS demand careful positioning and a reconfirmation of appropriate ETT position, airway pressures and lung compliance is recommended in these children.

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## 1. Klippel Feil Syndrome

The classic clinical triad of a short neck, a low posterior hairline, and severe restriction of neck movements due to complete fusion of the cervical spine forms the hallmark of Klippel-Feil syndrome (KFS) which was first reported in 1912 by Klippel and Feil.<sup>1</sup> Two overlapping classification systems exist for KFS. The original classification of KFS subtypes (I, II and III) described by Maurice Klippel and Andre Feil<sup>1</sup> and one more recent updated classification of KFS Classes (KF1-4) as described by Clarke and colleagues<sup>2</sup> [Clarke et al 1998]. The original classification differentiate individuals based on the degree of cervical fusion; KFS type I is characterized by extensive fusion of vertebrae of the neck (cervical vertebrae) and the upper back (i.e., upper thoracic vertebrae); type II is characterized by fusion at one or two cervical or thoracic vertebrae; type III is characterized by fusion of vertebrae of the neck as well as vertebrae of the upper or lower back (i.e., lower thoracic or lumbar vertebrae). In contrast, the classification described by Clarke et al<sup>2</sup> differentiate individuals based

on patterns of inheritance, common associated anomalies and the axial level of the most anterior fusion. KF1 is the only class presenting with C1-2 fusion, the very short neck and recessive inheritance; KF2 is autosomal dominant with the most anterior fusion at C2-3; KF3 is recessive or has reduced penetrance of isolated fusions between any of the cervical vertebrae except C1-2; KF4 includes cases of Wildervank and Duane syndrome. In most individuals with KFS, the condition appears to occur randomly for unknown reasons (sporadically). However, in other cases, familial patterns have been reported that indicate autosomal dominant or autosomal recessive inheritance. Here we are reporting a case of paediatric KFS and reviewing the anaesthetic concerns and management.

## 2. Case Report

A five year old boy weighing 15 kg was referred by the orthopaedics department for repair of Sprengel deformity. A preanaesthetic assessment revealed uneventful perinatal period & no history of cardiovascular or respiratory insufficiency. On examination, the child had a small atretic encephalocele at the posterior fontanelle, a short webbed

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neck, low posterior hair line, Sprengel's deformity in right shoulder (Figure 1) and scoliotic deformity of the spine with convexity towards left side. Airway examination showed adequate mouth opening, Modified Mallampatti Class (MMPC) 2, Upper Lip Bite Test (ULBT) grade 2 & complete inability to flex or extend the neck. Computerized tomography (CT scan) and magnetic resonance imaging (MRI) spine showed atlanto-axial subluxation with retrodens spinal canal stenosis and fusion of the C4-C5 & C6 vertebrae.



**Fig. 1:** A photograph of a child Sprengel's deformity in right shoulder

We used ASA difficult airway algorithm to formulate our airway management plan to manage this difficult airway (lack of cooperation, limited neck movements and associated malformations of laryngeal cartilages.)<sup>3</sup> Our plan A was check laryngoscopy under general anaesthesia with manual in line stabilization (MILS), assessing for ability to control ventilation, followed by muscle relaxation and endotracheal intubation if the laryngoscopic view was Cormack Lahane grade (CL Grade) 1 or 2 and controlled ventilation was possible. If controlled ventilation was not found to be easy, but laryngoscopic view was good, the intubation was to be done without muscle relaxation preserving spontaneous breathing (plan B). In the event of CL grade being 3 or more, Fiberoptic guided intubation under general anaesthesia with spontaneous breathing preserved as plan C. Placement of supraglottic airway was

considered as the rescue plan.

After the patient was brought into the operating room para- oxygenation was initiated with nasal prongs and preoxygenation with 100% oxygen was done using face mask, till FeO<sub>2</sub> was more than 90%. 0.25 mg midazolam & 30mcg of fentanyl were administered intravenously. Anaesthesia was induced using 2 mg/kg of propofol, mask ventilation checked, found to be easy, following which a check laryngoscopy was done with MILS. Visualization of the vocal cords was achieved with ease, with CL grade 2. The patient was then paralyzed with 0.1 mg/kg vecuronium and endotracheal intubation with tube size 5 mm was performed. MILS was maintained and para- oxygenation provided throughout the airway management. ETT's position was confirmed clinically and by capnography.

The patient had a component of scoliosis and decreased air entry on the left side was comparable to the pre op decreased air entry, with peak airway pressures between 10- 12 cm of H<sub>2</sub>O which was satisfactory. So, we decided to proceed with the prone positioning. Any unwanted movement at the neck were prevented and the patient was moved with all body parts aligned in anatomical position. After positioning we re- confirmed the positioning of the tube. When surgeons started to paint and drape the patient, the peak airway pressures rose to 30 cm of H<sub>2</sub>O while plateau pressure was still 15 cm of H<sub>2</sub>O. The EtCO<sub>2</sub> value at that point was 42 cm of H<sub>2</sub>O with an obstructive upsloping pattern. Air entry decreased considerably bilaterally. The abdomen was free from bolsters but the chest expansion had considerably decreased. We increased the depth of anaesthesia by increasing the sevoflurane concentration but it had no effect. Since the neck of the kid was short and muscular, the sandbag under the chest started compressing the pliable trachea and the ventilation was hampered. Once the bolsters were properly re-positioned, normal ventilation was resumed with appropriate airway pressures. At the end of the surgery the child was reversed and extubated uneventfully, while minimizing neck movements.

### 3. Discussion

KFS is a bone disorder occurring due to failure of normal segmentation of the cervical somites in the 3rd to 8th week of gestation. Our patient had type 3 KFS with fusion of the cervical vertebrae along with thoracic & lumbar spine scoliosis. Associated anomalies were an encephalocele with atlanto-axial instability.

Patients with KFS pose numerous problems to the anaesthesiologist. They have cervical instability and are at high risk for spinal cord injury during laryngoscopy, intubation and positioning for surgery.<sup>4</sup> In 1988, Daum and Jones<sup>5</sup> suggested that the most prudent and effective way is an awake fiberoptic intubation. The nasal route is preferred since the tongue is out of the way and the patient cannot bite

down on the tube or scope. However in paediatric patients oral intubation with an appropriate oropharyngeal airway may be preferred as adenoid injury is expected in this patient population. Use of cricoid pressure in patients with unstable cervical spine is controversial as it can potentially lead to cervical subluxation.<sup>6</sup> The intubating LMA has also been used to facilitate intubation without manipulation of the head and neck. Keller and Brimacombe<sup>7</sup> suggest that cervical pressures generated by the Laryngeal mask devices can produce posterior displacement of the normal cervical-spine. Therefore caution must be used when extrapolating these findings to the unstable cervical-spine.

Apart from airway related difficulties, cardiovascular anomalies have been recognized in patients with KFS<sup>8–10</sup> with an incidence of 4.2 to 14 per cent which warrants a thorough cardiovascular examination. Significant scoliotic deformity leads to restrictive lung disease due to lung tissue compression. Pulmonary hypertension progressing to cor pulmonale or shunt reversal in case of a co-existing VSD are prominent causes of mortality in this population. Due to the restrictive nature of the disease, the patients have decreased vital capacity which manifests as dyspnea on exertion. Our patient had none of these symptoms.

Prone positioning in these patients also places these patients at an additional risk. As for all patients placed prone under GA, it has to be made sure that limbs are padded and are in neutral position to avoid impingement and excessive stretch forces. Head may be turned to side or facedown making sure that weight is borne by bony structures only and no soft tissue is compressed. In addition, the asymmetric neck and thorax may require careful positioning of the sand bags preventing any increased intra- abdominal & intra thoracic pressures, pressure on neck and ensuring that the trachea is not kinked. The ETT position, airway pressures, chest compliance as well as the delivered tidal volume must be rechecked after placing the patient prone.

#### 4. Conclusion

A careful assessment of airway is needed in children with KFS. Identification of available and difficult areas of airway along with an algorithmic approach helps to formulate the airway management plans and successfully execute them. The various spine deformities associated with KFS demand

careful positioning and a reconfirmation of appropriate ETT position, airway pressures and lung compliance is recommended in these children.

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None.

#### 6. Conflict of Interest

None.

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