ANAESTHETIC CHALLENGES IN A PATIENT WITH KLIPPEL FEIL SYNDROME UNDERGOING SURGERY

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SUMMARY
A 37 year old male with Klippel Feil Syndrome was scheduled for laparoscopic cholecystectomy and left inguinal hernioplasty. We present the anaesthetic management of this patient highlighting the various anomalies associated with Klippel Feil Syndrome and the presence of a difficult airway.

Keywords : Klippel Feil Syndrome, Difficult airway.

Introduction
Klippel Feil Syndrome was described independently in 1912 by Maurice Klippel and Andre Feil. They described patients who have a short neck, decreased range of motion in the cervical spine and a low posterior hairline.

Feil subsequently classified the syndrome into three categories. Type 1 is described as extensive fusion of the cervical and upper thoracic spines. Type 2 is present when there are one or two interspace fusions often associated with hemi vertebrae and atlanto-occipital fusion. Type 3 occurs when thoracic and lumbar spine anomalies are associated with type 1 or type 2 Klippel Feil syndrome.

Case report
A 37 year old male presented with complaints of epigastric and right hypochondrial pain of six months duration which had worsened over the past three months. He also complained of a swelling in left groin since the past one year. He had four to five of such episodes of mild upper abdominal pain per day each episode lasting for five to ten minutes. There was no associated fever with chills, jaundice or vomiting. The patient also had a left inguinal swelling which increased on coughing and reduced spontaneously.

The patient had been diagnosed to have Klippel Feil syndrome several years previously and was a known diabetic on diet control. He was a non smoker, not a hypertensive and did not have asthma.

On examination the patient was a short, moderately built and nourished gentleman with a pulse rate of 85 / minute and a blood pressure of 130/80 mm of Hg. He had a short webbed neck Fig. 1 and a low posterior hairline Fig. 2 with no flexion or extension of the neck Fig. 3 and a small cystic swelling in the region of the occiput. He had several limb anomalies, on his right hand he had six digits, the right fifth metacarpal was absent and he had wasting of his right arm. He had bilateral pes cavus and short 4th and 5th metatarsals. Examination of the airway showed that there was no flexion or extension of the neck, he had adequate mouth opening and his Mallampatti grade was 3. The systemic examination was normal.

Fig. 1 : Webbed Neck
Fig. 2 : Low Posterior Hairline
Fig. 3 : Limited Extension
On investigation, haemoglobin, creatinine and electrolyte estimations were normal. He was found to have elevated blood sugar which were adequately controlled with a diabetic diet. He also had elevated serum cholesterol for which he was treated with HMG CoA reductase inhibitors. The ultrasound of the abdomen confirmed gall stones and also showed a left renal calculous. An electrocardiogram and chest x-ray was normal. Cervical spine x-rays done in flexion and extension Fig. 4 did not show instability of the cervical spine. There was fusion of the cervical spines from C2 to C7.

On the evening before surgery, the procedure was explained the patient. He was kept nil by mouth for 8 hours and medicated with diazepam 5 mg per oral 90 minutes prior to surgery.

The patient was brought into the anaesthesia room and an intravenous access secured with a 16 G canula, in a peripheral vein of his left hand under local anaesthesia. We then proceeded to anaesthetize his upper airway with local anaesthesia after first calculating the maximum dose of local anaesthetic that could be used. The patient was first given 5 ml of 2% xylocaine viscous gargles which he was allowed to swallow so as to coat the posterior surface of the epiglottis. This was followed by 3 aliquots of a 10% xylocaine spray which was instilled into the posterior oropharynx, with the patient taking deep breaths. Minimal sedation of 1mg of midazolam was given intravenously to keep the patient calm and glycopyrolate 0.2 mg was given to reduce secretions. His nose and nasopharynx was anaesthetized with single aliquots of 2% cophenalocaine spray in each naris. The patient was then shifted to the operating room where his ECG, SpO2, saturation and BP were monitored. He was induced with increments of fentanyl and propofol intravenously. It was found that the patient was difficult to ventilate and he tended to obstruct. An oral airway was inserted to help maintain the airway. When sufficient depth of anaesthesia was reached, a check laryngoscopy was done and there was difficulty in visualization of the vocal cords. The patient was allowed to wake up and it was decided to proceed with awake fibreoptic intubation.

A nasopharyngeal airway well lubricated with 3ml of 2% lidocaine jelly, which was slit longitudinally was inserted into the right nostril. A 7.5 mm cuffed endotracheal tube was threaded over the bronchoscope. The scope was then carefully maneuvered through the slit nasopharyngeal airway until the vocal cords were visualized. 2 ml of 2% lidocaine was injected through the side port of the fibreoptic bronchoscope, using the ‘spray as you go’ technique to anaesthetize the vocal cords. The fibreoptic bronchoscope with the endotracheal tube threaded over it was then introduced into the trachea, the tracheal rings and the carina identified. Propofol 20 mg was given intravenously, the endotracheal tube inserted and its position confirmed after removal of the fibreoptic bronchoscope, by the chest movements as seen on inspection and bilateral equal air entry on auscultation. The EtCO2 further confirmed correct placement of the endotracheal tube. The patient was then paralyzed with a non-depolarizing neuromuscular blocker and anaesthesia was maintained with halothane, 66% nitrous oxide and 33% oxygen. IPPV was established with a Datex Ohmeda 7000 series ventilator.

Surgery proceeded uneventfully and residual neuromuscular blockade was reversed with atropine 0.02 mgkg⁻¹ and neostigmine 0.05 mgkg⁻¹. The tracheal tube was removed after resumption of spontaneous breathing and the return of good muscle tone. Post-operatively the patient was monitored in the recovery room after which he was sent to the ward.

**Discussion**

Patients with Klippel Feil syndrome present at different ages with varying clinical presentations. This syndrome may also be detected as an incidental finding. The incidence of Klippel Feil syndrome has been investigated in 2 studies. Gjorup reviewed all cervical spine films in a single hospital in Copenhagen and determined an incidence of 0.2 cases per 1000 people. Brown reviewed 1400 skeletons...
from the Terry Collection at Washington University School of Medicine and he found an incidence of 0.71%. It occurs due to a failure of normal segmentation of the cervical somites in the 3rd to 8th week of gestation. Its etiology is unknown.

In addition to the upper cervical involvement, patients also present with facial asymmetry and torticollis which occurs in 21-50% of patients. Orthopaedic and neurological manifestations are seen in 20% of patients of which occipitocervical abnormalities are the most common. Scoliosis occurs in approximately 60% of patients. A Sprengel’s anomaly occurs in 20-30% of patients. An omovertebral bone, an osteocartilagenous connection, that tethers the scapula to the spine, may present as decreased range of movement of the shoulders. Some patients may present with synkinesia. Renal anomalies are common in individuals with Klippel Feil syndrome, including a double collecting system, renal ectopia and bilateral tubular ectasia. Major renal anomalies include hydronephrosis, absence of a kidney and a horse shoe kidney. Hearing loss is common with Klippel Feil syndrome and can be sensorineural, conductive or mixed. Cardiovascular anomalies occur in 14-29% of patients, the most common being Ventricular Septal Defect. Other less common anomalies are congenital limb deficiencies, craniosynostosis, ear abnormalities and craniofacial abnormalities. It can also present with a variety of other clinical syndromes like, foetal alcohol syndrome, Goldenhar syndrome and other anomalies.

Our patient had type 1 Klippel Feil syndrome with massive fusion of the cervical spines. Associated anomalies were a dermoid cyst in the occiput, polydactyly and clinodactyly of his right hand and pes cavus with absent 4th and 5th metatarsals bilaterally. He did not have any atlantoaxial instability.

These patients have a potentially unstable cervical spine and abnormal atlanto-occipital junction and are prone to an increased risk of neurological damage. Syncopeal attacks may be precipitated by sudden rotatory movements of the neck in patients with Klippel Feil syndrome. O’Conner and Moysa reported that airway control can be temporarily lost after induction and an LMA may be required to attain control of the airway. There was also a failure in fibreoptic intubation on two separate occasions. The proposed surgery of the patient was cancelled twice due to failure to intubate.

These patients present to us for several reasons, such as, correction of associated anomalies like scoliosis, spinal canal stenosis, renal anomalies etc. They also present with other unrelated problems as our patient did with chronic calculous cholecystitis and a right inguinal hernia. Patients with Klippel Feil syndrome pose a challenge to the anaesthetists with regard to the management of the difficult airway.

Anaesthetists responsible for securing the airway in patients with an abnormal cervical spine have several options. In 1988, Daum and Jones suggested that the most prudent and effective way is an awake fibreoptic intubation with good conditions found in awake patients since they can assist in clearing their own secretions, phonating or panting. 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 oral intubation may be attempted using an appropriate airway cum bite block like the ovassapian airway. Blind nasotracheal intubation is an alternate mode of securing the airway in difficult intubations and in those with cervical spine injury. This technique however is associated with flexion and extension, two maneuvers which are avoided in patients with an unstable spine. Anterior neck pressure is also given to stabilize the larynx. This technique has a failure rate of 3-20% while traumatic intubation soiling the airway is reported as 7-8%. The use of cricoid pressure in patients with unstable cervical spines is also controversial as there is the potential for subluxation at the site of instability following application of the cricoid pressure.

The intubating LMA has also been used to facilitate intubation without manipulation of the head and neck. Keller and Brimacombe 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.

Taking all the modes of airway management into consideration, we feel that the optimum mode of intubation in a patient with cervical spine pathology is an awake fibrescope aided intubation. The advantages are, (1) an awake spontaneously breathing patient who is maintaining his own airway (2) no spinal movement is needed during intubation, (3) a tool for intubation that allows confirmation of tracheal tube placement, (4) it has a high rate of success (5) low rate of complications and (6) good patient acceptance.

A difficult airway must be approached with caution. A comprehensive preoperative examination and ‘work up’, the availability of several alternate techniques, a willingness to call an expert help, surgeons standing by to provide a surgical airway and/or moral support and a good deal of common sense go a long way in ensuring a favorable outcome.
References


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