

Anaesthetic challenges in a patient with Klippel–Feil syndrome scheduled for panendoscopy and biopsyⁱ

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ⁱWritten consent was obtained from the patient

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Klippel–Feil syndrome is one of the congenital causes of difficult airway. It is characterised by a classic triad of a short neck, restricted cervical spine movement, and a low posterior hairline, which can pose a significant challenge to the anaesthetist during airway management. A case of Klippel Feil Syndrome type 2 with associated Sprengel's deformity for panendoscopy under general anaesthesia is presented. The anaesthetic considerations in the management of this patient are also discussed.

Keywords: awake-fibreoptic intubation, difficult airway, Klippel-Feil syndrome, panendoscopy, Sprengel's deformity

Case report

We present a 41-year-old male who was referred to the Ear, Nose and Throat (ENT) clinic with a two-month history of worsening dysphagia, hoarseness and choking. At the ENT clinic a flexible nasendoscopy revealed an exophytic lesion on the epiglottis. The vocal cords were not visible. An urgent panendoscopy and biopsy under general anaesthesia was therefore scheduled by the ENT surgeon to ascertain the underlying pathology of the lesion.

The patient was a known case of Klippel–Feil syndrome with a Type II variant associated with Sprengel's deformity. His other medical history included spina bifida affecting the thoracic spine, unilateral renal agenesis and previous corrective heart surgery as a child for congenital heart disease. He took amlodipine to control his hypertension.

Physical examination demonstrated a height of 165 cm, weight of 76 kg and a BMI of 28 kg/m². Airway examination revealed a small mouth opening and a Mallampati class IV (Figure 1). He was noted to have a very short webbed neck with a thyromental distance of 1 cm, no flexion–extension, and limited sideways neck movements (Figure 2).

Systemic examination revealed no further abnormalities.

Routine blood investigations, electrocardiogram, pulmonary function tests, and chest X-rays were all within normal limits, and a neurological assessment was also normal.

Computed tomography (CT) of the neck and thorax with contrast revealed an irregular space-occupying lesion measuring 3 x 2.5 cm at the left pyriform sinus involving the left aryepiglottic fold and epiglottis. Anteroposterior and lateral cervical spine X-rays together with a CT cervical spine demonstrated fusion of the upper cervical (C2–C7) vertebrae, an omovertebral bone and spinal bifida, suggestive of Sprengel's deformity (Figure 3).

Following a multidisciplinary team meeting, an awake video-assisted fibre-optic intubation using an Ambu® aScope 3 (Ambu A/S, Ballerup, Denmark) was considered the most appropriate anaesthetic choice. Thus, our anaesthetic airway management options included awake video-assisted fibre-optic intubation

using the Ambu® aScope 3 and utilising the GlideScope® (Verathon Inc, Bothell, WA, USA) as an alternative. We discussed these options with the patient after the multidisciplinary team meeting.

Following the establishment of standard monitoring and intravenous access, remifentanyl target-controlled infusion effect-site concentrations of 1–2 ng/ml were started. A bolus dose of 1 mg midazolam was also given.

We calculated the maximum safe dose of lignocaine at 9 mg/kg, based on lean body weight for topical anaesthesia in adults. Co-Phenylcaine Forte was administered via a mucosal atomiser device (MAD) to both nostrils to anaesthetise the nasopharynx. Oxygen at 4 l/min was also given through the opposite nostril using a nasal cannula.

Co-Phenylcaine via mucosal atomiser device (MAD) was administered to both nostrils; 10% lignocaine was used to anaesthetize the back of the throat. A 6.0 mm cuffed endotracheal tube was threaded over the Ambu® aScope with a 16-gauge epidural catheter advanced through the working channel of the scope.

After multiple unsuccessful attempts in the semi-recumbent position we decided to change the patient's position into a sitting upright position. At this position we were able to visualise the vocal cords. On visualisation of the vocal cords, 1–2 ml of 2% lignocaine was used as spray-as-you-go technique. The scope with endotracheal tube was then introduced into the trachea. Propofol 150 mg was given intravenously with the endotracheal tube secured and its current position confirmed with an end-tidal carbon dioxide monitor. A non-depolarising agent was given intravenously and anaesthesia was maintained using sevoflurane in air and oxygen via intermittent positive-pressure ventilation. Ondansetron 4 mg, dexamethasone 3.3 mg and paracetamol 1 g were all administered intravenously as routine intraoperative medications. A summary of our awake video-assisted fibre-optic intubation technique is given in Table 1.

Surgery and anaesthesia proceeded uneventfully. At surgery, a large exophytic lesion arising from the aryepiglottic fold was exposed. This mass was debulked and a biopsy was obtained. At the end of surgery the residual neuromuscular blockage was



Figure 1: Showing a short webbed neck with Mallampati grade IV.



Figure 2: Showing the Sprenkel's deformity and spina bifida with severely restricted neck extension and flexion.

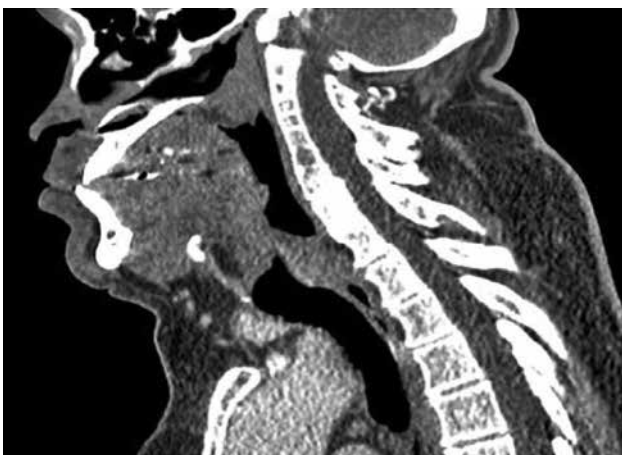


Figure 3: Lateral cervical spine CT in extension.

then reversed and the patient was successfully extubated fully awake following DAS extubation guidelines.

We electively monitored the patient in our recovery room for an extended period of time and the patient was discharged the same day having fully met the day unit discharge criteria. Following postoperative multidisciplinary team meeting review, the patient has commenced chemo-radiotherapy with curative intent.

Discussion

Maurice Klippel and Andre Feil first described Klippel–Feil syndrome independently in 1912.¹ Klippel–Feil syndrome is one of the congenital abnormalities associated with an anticipated difficult airway, and has an estimated incidence of 1:40 000 to 1:42 000 births.² It is an inherited skeletal disorder that occurs as a result of abnormal segmentation of the cervical somites during organogenesis between the third and eighth week of gestation.³

Four variants of Klippel–Feil syndrome have been described.⁴ Table 2 summarises the characteristic features of these four variants.

Klippel–Feil syndrome patients often have an unstable cervical spine and atlanto-occipital fusion, making them prone to neurological damage during laryngoscopy, intubation, patient transfer and extubation.⁵ Although in our case neurologic assessment was normal, we took all necessary neck precautions during patient intubation, positioning and extubation.

Klippel–Feil syndrome has also been associated with other anomalies as indicated in Table 1. Our patient had Sprengel's deformity and kyphoscoliosis, which can both potentially pose difficulties in patient positioning. These features may have contributed to the multiple failed awake fibre-optic intubation attempts in our patient's initial semi-recumbent position. Additionally, the presence of these features may have compromised our patient's ventilation thus causing a risk of hypoxia, especially considering our patient was undertaking airway surgery.⁶ To our knowledge, this is the first case report of a panendoscopy in a Klippel–Feil syndrome patient requiring a general anaesthetic.

An awake video-assisted fibre-optic intubation is an essential skill in the management of a patient with an anticipated difficult airway, and has been previously utilised for cases of Klippel–Feil syndrome^{7–9} and other difficult airway management.^{10,11} Airway assessment and radiological imaging of our patient predicted a difficult airway and thus we electively decided to utilise awake fibre-optic intubation using the Ambu® aScope 3. We initially envisaged difficulties in nasal awake fibre-optic intubation due to the very narrow anterior nasal nares, and thus we decided to employ a GlideScope® awake fibre-optic intubation approach via the oral route. Unfortunately, we were unable to insert the GlideScope® into the mouth due to the size of the patient's tongue and his very short thyromental distance. The Italian Society of Anaesthesia, Resuscitation and Intensive Therapy (SIAARTI) recommend the use of a video laryngoscope only in predicted difficult airway in patients with adequate mouth opening.¹² A nasal awake fibre-optic intubation approach is instead favoured as this ensures that the tongue is out of the way and the patient cannot bite on the scope.⁶

Given these challenges, awake fibre-optic intubation is the recommend approach to ensure a safe and secure airway.^{13,14} This approach does not require any spinal movement and allows for an awake spontaneously breathing patient who can maintain his/her own airway and can assist in clearing of his/her own

Table 1: Summary of our technique for awake video-assisted fibre-optic intubation

Local anaesthetic technique	Sedation	Instruction to patient
Nose: • 0.5 ml Co-Phenylcaine spray via MAD to each nostril	Midazolam 1 mg Remifentanil TCI 1–2 ng/ml	Reassured the patient
Pharynx: • Lignocaine 10% spray to tip of tongue • 4 sprays to superior pharynx • 4 sprays to inferior pharynx	Remifentanil TCI 1–2 ng/ml	Reassured the patient Swallow
Larynx: • Lignocaine 2% spray via the epidural catheter above the cord and below the cord (2 ml each)	Remifentanil TCI 1–2 ng/ml	Deep breath Deep breath Reassured and encouraged
Intubation: • Ambu aScope ³ passed through the cords and the ETT passed and secured	Propofol	Deep breath Reassured and encouraged

Notes: MAD: mucosal atomisation device, TCI: target-controlled infusion, ETT: endo-tracheal tube.

Table 2: Summary of variants of Klippel–Feil syndrome (KFS).

KFS type	Features
Type I	Extensive fusion of cervical and upper thoracic vertebrae into a single block
Type II	Failure of complete segmentation occurs at one or two cervical interspaces and hemivertebrae Most common type
Type III	Includes Type I or Type II features May involve coexisting fusion in the lower thoracic and lumbar spine
Type IV	Klippel–Feil anomaly associated with sacral agenesis
Other features	Kyphoscoliosis, torticollis, renal anomalies, Sprengel's deformity, congenital heart disease, synkinesia, hearing loss

secretions. Moreover, it has a high rate of success, low rate of complications and good patient acceptance.¹⁴

Conclusion

A difficult airway must be approached with meticulous attention to detail. Our report highlights the importance of robust patient information, thorough perioperative evaluation and a detailed airway management plan as keys to successful airway management in this group of patients. Awake fibre-optic intubation is considered the most prudent and effective way to manage an airway in a Klippel–Feil syndrome patient.

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