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

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Written 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-fiberoptic 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.

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Physical examination demonstrated a height of 165 cm, weight of 76 kg and a BMI of 28 kg/m². Airway examination revealed a short neck with 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. Anterioposterior 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 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, remifentanil 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 for anaesthetise the naso-oropharynx. Oxygen at 4 l/min was also given through the opposite nostril using a nasal cannula.

Co-Phenylicaine 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

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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
don 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
syndrome5–8 and other difficult airway management.9,10 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.11 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.12,13 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

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

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

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

Figure 3: Lateral cervical spine CT in extension.
manage an airway in a Klippel–Feil syndrome patient. intubation is considered the most prudent and effective way to management in this group of patients. Awake fibre-optic airway management plan as keys to successful airway information, thorough perioperative evaluation and a detailed to detail. Our report highlights the importance of robust patient A difficult airway must be approached with meticulous attention complications and good patient acceptance. 14

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

<table>
<thead>
<tr>
<th>Local anaesthetic technique</th>
<th>Sedation</th>
<th>Instruction to patient</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nose:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>- 0.5 ml Co-Phenycaine spray via MAD to each nostril</td>
<td>Midazolam 1 mg</td>
<td>Reassured the patient</td>
</tr>
<tr>
<td>Pharynx:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Lignocaine 10% spray to tip of tongue</td>
<td>Remifentanil TCI 1–2 ng/ml</td>
<td>Reassured the patient</td>
</tr>
<tr>
<td>- 4 sprays to superior pharynx</td>
<td>Remifentanil TCI 1–2 ng/ml</td>
<td>Swallow</td>
</tr>
<tr>
<td>- 4 sprays to inferior pharynx</td>
<td>Remifentanil TCI 1–2 ng/ml</td>
<td>Swallow</td>
</tr>
<tr>
<td>Larynx:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Lignocaine 2% spray via the epidural catheter above the cord and below the cord (2 ml each)</td>
<td>Remifentanil TCI 1–2 ng/ml</td>
<td>Deep breath</td>
</tr>
<tr>
<td>Intubation:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Ambu aScope® passed through the cords and the ETT and passed and secured</td>
<td>Propofol</td>
<td>Deep breath</td>
</tr>
<tr>
<td>Notes: MAD: mucosal atomisation device, TCI: target-controlled infusion, ETT: endo-tracheal tube.</td>
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Table 2: Summary of variants of Klippel–Feil syndrome (KFS).

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

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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References