Case Reports

ANESTHETIC MANAGEMENT IN A CASE OF KLIPPEL-FEIL SYNDROME AND LITERATURE REVIEW

ZAHID HUSSAIN KHAN, M.D., HASSAN REZA KHODADADI, M.D., AND PAYAM EGHTESADI-ARAGHI, M.D.

From the Department of Anesthesiology, Imam Khomeini Hospital Complex, Tehran University of Medical Sciences, Tehran 14197, Iran.

ABSTRACT

Klippel-Feil syndrome is known by the classic triad of shortness of the neck, limitation of neck movements, and a low posterior hairline. There are often accompanying cervical spinal abnormalities such as kyphoscoliosis as well as urogenital and cardiac abnormalities.

Presented here we have a 20 year old young man with hypoesthesia and decreased motor function in the right hand. The problem began one year back following a minor head trauma and had a progressive course involving the legs, especially the feet. Cervical magnetic resonance imaging was compatible with C3-C4 cord compression as well as blocked vertebrae. The patient was evaluated to be in Mallampati class II. Endotracheal intubation was performed employing gentle manual axial traction in both anterior and posterior operative approaches without any neurological sequela. It is recommended that in situations where fiberoptic or Bullard laryngoscopes are not available and Mallampati class is low, direct laryngoscopy associated with gentle axial traction may be a plausible substitute.

INTRODUCTION

Klippel-Feil syndrome was described for the first time by Klippel and Feil in 1912. The syndrome is associated with short neck and limitation in cervical movements. It is observed in 0.2 to 4.2 neonates out of 1000 live births. In some patients, cervical spine abnormalities associated with kyphoscoliosis, and urogenital abnormalities and cardiac anomalies are observed.

These patients may show neck rigidity and limited neck extension due to vertebral fusion, making endotracheal intubation a difficult and potentially hazardous task. The patients are susceptible to spinal cord injuries and neurological deficits after laryngeal intubation with recommendations by some to avoid induction of general anesthesia by intravenous agents in these particular patients if possible.

However in patients in whom general anesthesia is warranted, various methods such as awake intubation, fiberoptic laryngoscopy, Bullard laryngoscopy, cricothyroidotomy and transtracheal ventilation have been suggested. Airway examination in our case revealed...
the soft palate and the base of the uvula, but not the
tonsillar faucets (Mallampati class II).

Our patient was intubated twice for two different sur-
gical approaches under general anesthesia, employing
axial in-line traction and direct laryngoscopy. The intu-
bation had been relatively easy and uneventful.

CASE REPORT

A twenty year old man complaining of mild weakness
and paresthesia in all the four limbs was admitted at this
center in Sept. 2002 for operative treatment of the cervi-
cal spine. The illness started one year back and had a
progressive course necessitating his admission. There
was also a positive history of minor head trauma at the
onset of the symptoms. Plain radiographs revealed fu-
sions at C3 to C4 levels (Fig. 1), and magnetic resonance
imaging showed C3-C4 disc herniation and canal steno-
sis at the same level causing severe cord edema (Fig. 2).
A positive family history for such conditions was not
forthcoming. The patient had a low hairline associated
with neck deformity and an apparent cervical scoliosis
(Fig. 3). There was limited neck extension (≤50°) associ-
ated with a positive Lhermitte’s sign. Flexion and rota-
tion were however not limited. Airway examination re-
vealed Mallampati class II. Thyromental span was 8 cm
and normal. No dental abnormality or occlusion was
present. Cardiopulmonary and genitourinary evaluation
proved to be normal.

General anesthesia for cervical laminectomy and pos-
terior fusion was performed employing a balanced stan-
dard anesthesia under conventional monitoring devices.
Prior to induction, diazepam 7.5 mg and fentanyl 100 µg
were administered. Induction was conducted using thi-
pental sodium 300 mg and succinylcholine 80 mg was used
to achieve muscular relaxation. Lidocaine 90 mg was in-
jected soon after succinylcholine to ensure hemodynamic
stability. The head traction was applied gently by an
attending neurosurgeon and laryngoscopy conducted
with a Macintosh blade 3 (Riester, Germany) employing
a No. 8 cuffed Roche(™) oro tracheal tube. At laryngos-
copy the posterior extremity of the glottis was visible
and intubation was successful during the first attempt.
After securing the orotracheal tube, the patient was ro-
tated to the prone position with necessary precaution
and maintenance of anesthesia managed with halothane

![Fig. 1. A plane radiogram revealing C3-C4 instability associated
with blocked vertebrae at superior and inferior levels.](image)

![Fig. 2. Magnetic resonance weighted image of cervical spine re-
vealing C3-C4 disc herniation associated with narrow canal at the
same level. Note increased signal intensity in the spinal cord in
favor of cord edema.](image)
6%, a 50% mixture of N₂O and O₂ along with incremen­
tal doses of pancuronium bromide and fentanyl. After 
completion of the surgery, which lasted 120 min, the pa­
tient was returned back to the supine position and re-
duced muscle paralysis reversed with neostigmine 2.5mg
and atropine 1.2mg. Extubation of the trachea was ac-
complished after ensuring that the patient was awake
and had adequate tidal respiration. The same anesthetic
protocol was again repeated 14 days later for anterior
discectomy and fusion with no sequela except mild sore
throat.

DISCUSSION

Patients with Klippel-Feil syndrome are prone to ma-
nor neurological sequela following minor trauma due to
spinal spine instability. C₂-C₃ and C₃-C₄ are the most
commonly involved interspaces. However multisegment
involvement is not uncommon. Our patient had his C₂-
₃ disc space involved.

General anesthesia may become a necessity for these
patients and a thorough physical examination is essen-
tial to unveil any potential risk. The patients pose the
anesthetist to a high risk of difficult intubation as well
as spinal cord damage due to instability. Manipula-
tion of the neck or an attempt to extend the neck during laryn-
goscopy and thereafter must be carefully controlled if
neurological damage is to be avoided.

Despite the afore mentioned problems encountered
during intubation, there still does not exist a formal con-
sensus on an intubation technique in these cases. Sev-
eral options such as awake laryngoscopy under local anesthesia, inhalation induction with halothane, inser-
tion of a laryngeal mask airway (LMA) and oral fiberoptic intubation (FIB), have been advocated as possible alternatives in patients with Klippel-Feil syn-
drome. The most commonly advocated technique in these cases has been flexible fiberoptic laryngoscopy. The

technique is safe but difficult at times and entails some
time. At times Bullard laryngoscopy has been proposed
to be safe, however it is not always available and re-
mains ubiquitous in operative room settings. Awake in-
tubation may be a safe method but it may increase in-
tracranial pressure and furthermore there is limited view
for the anesthetist. Cricothyroidotomy is considered to be
a good choice but extubation remains a dilemma and
therefore patients might have massive subcutaneous emphysema or cause airway injury as

In our patient, laryngoscopy was performed under axial-in-line manual traction. During the first operation,
intubation could be performed on the first attempt, but
during the second operation, the intubation was suc-
cessful after a second attempt and light pressure on the larynx was needed to accomplish the task. The patient
on laryngoscopy had a Cormack and Lehane grade which did not cause much of a problem during the first
attempt, but caused some difficulty during the second
time. The difficulty during the second time could be at-
tributed either due to the cervical laminectomy and pos-
terior fusion performed during the first stage which had
made laryngoscopy difficult or perhaps an extra caution
on our part to avoid cervical spinal cord damage.

Because of predicted difficulty with tracheal intuba-
tion in patients with Klippel-Feil syndrome, it is indis-
penable to secure the airway before induction of gen-
eral anesthesia especially in emergent cases because
emergency endotracheal intubation is normally consid-
ered to be difficult because of the altered anatomy and
reduced neck mobility. In our patient the intubation
was successful because earlier prediction of the airway
was found to be easy, the lesion level was at C₄-C₅ interspace away from the atlanto-occipital region and above
all the procedure was cautiously accomplished under axial traction. Although FIB has been recommended in
such cases, this modality might fail, cause massive subcutaneous emphysema or cause airway injury as
reported by Maktabi et al. in three of their cases. It could however be argued that if the scenario had progressed to a "can’t intubate - can’t ventilate" situation in this particular patient after having been paralyzed, and intubation had proved impossible, what would have been the plan B in our minds. Under such circumstances, we would have opted for an intubating laryngeal mask airway (ILMA) since an overall success rate of 100% has been reported with it. A similar success rate has also been reported with FIB, but oxygen desaturation occurred more frequently with it.

Our patient was to undergo an elective surgical procedure which provided us with ample time to assess the airway and thus enabled us to opt for a method that would not only prove to be safe for the patient but at the same time would be easy to accomplish.

Finally, it is recommended that where fiberoptic or Bullard laryngoscopes are not available and airway class seems promising and easy, direct laryngoscopy associated with cautious manual axial traction under general anesthesia might be a plausible alternative in such cases.

REFERENCES