

REVERSE LMA INSERTION IN A NEONATE WITH KLIPPEL-FEIL SYNDROME

- Case report -

TARIQ AL ZAHRANI*

Klippel-Feil syndrome (KFS) was first described by Maurice Klippel and Andre Feil in 1912 in a patient with congenital fusion of cervical vertebrae¹. KFS is a complex syndrome of osseous and visceral anomalies that include the classical clinical triad of short neck, limitation of head and neck movements and low posterior hairline². It is associated with several defects, such as deafness, either conductive or neural, congenital heart defects, the most common being a ventricular septal defect, mental deficiency, cleft palate, rib defects, the Sprengel sequence (elevated scapula), and scoliosis. Patients with KFS exhibit a smaller lower third of the face and facial asymmetry with no dental implications. KFS occurs in 1 of every 42,000 births, and 60% of cases are in females².

We report a case of KFS with difficult airway using reverse LMA insertion technique.

Case Report

A new born 2 days old, female 2000g weight, 31 cm height was admitted to neonatal intensive care unit (NICU) after normal vaginal delivery. Examination revealed short webbed neck with restricted movement and low dorsal hair line, low set rotated malformed ear, high arched palate, narrow thoracic cage, and spinal deformity as shown on plain X-ray Fig. (1, 2). There was positive history of Klippel-Feil syndrome with one sibling. Echocardiography revealed small patent

* Address for correspondence: Tariq A. Al Zahrani, K.S.U.F.U, Tutor, Department of Anesthesia & ICU, College of Medicine, King Saud University, Riyadh, KSA. E-mail:tariq0404@hotmail.com.

ductus arteriosus defect (PDA). His plain X-ray revealed spinal vertebrae deformity. She was scheduled to have a diagnostic MRI under general anesthesia. As she had provisional diagnosis of KFS, the possibility of difficult intubation was entertained.

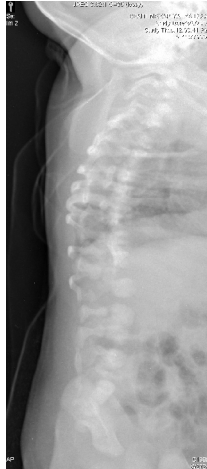


Fig 1
Condensed vertebrae, narrow thoracic cage, and spinal deformity



Fig 2
Short webbed neck with high arched palate

Anesthesia and monitoring equipment with non-ferromagnetic component was checked before providing anesthesia. She was monitored with ECG, pulse oxymeter, ETCO₂, non-invasive blood pressure and respiratory rate monitor in the radiology ward.

Anesthesia was induced with inhalation of sevoflurane gradual increment until adequate depth of anesthesia was obtained as judged by jaw relaxation. In order to avoid traumatization, LMA (size 1) was inserted with the reverse technique. LMA was held at the proximal end, insertion was conducted with the cuff fully deflated, facing the palate, and then rotated anticlockwise through 180 degrees as it was pushed into the hypopharynx. The cuff was inflated with 2-4 ml. air. The LMA was fixed after confirmation of its proper position (chest wall inflation, ETCO₂ and

auscultation). The patient breathed spontaneously throughout the procedure and she was monitored through closed TV circuit outside the MRI room displaying ECG, pulse oxymeter, ETCO₂ and respiratory rate. The procedure lasted 60 min. At the end, LMA was removed when she was fully awake, with no incidence of airway obstruction coughing, or laryngospasm.

Discussion

Endotracheal intubation in infants with Klippel-Feil syndrome may sometimes be impossible to accomplish with conventional methods. To aid difficult tracheal intubation many different techniques have been described. Moreover, securing airway and endotracheal intubation in patients with difficult airway especially in remote location like MRI suite, presents a real challenge and adds more difficulty to the anesthesiologist. In one case report³, it was reported that a guide wire inserted through LMA may be a successful alternative for the anesthesiologist dealing with difficult intubation in remote location anesthesia.

The LMA can be inserted in one of the two ways; either in the standard fashion, with the aperture facing anteriorly, according to the manufacture's instructions, or in reverse with the aperture facing the roof of the mouth turning the LMA through 180 degrees on reaching the posterior pharyngeal wall. The reverse technique has been selected because patients with KFS have atlantoaxial subluxation with the possibility of spinal cord injury with head extension. In addition, the reverse technique is easier for insertion. Once the LMA is inserted, the cuff is inflated with 2-4 ml air⁴. Both techniques provide similar success rates in achieving a clinically good airway⁵. In a report on the insertion of LMA⁶, it was reported successful in more than 90% of neonates on the first attempt which is consistent with the findings among adults. Difficult tracheal intubation in children and neonates is an important issue in anesthesiology and intensive care medicine. Down's syndrome, Klippel-Feil syndrome, Pierre Robin syndrome, Treacher Collins syndrome, mucopolysaccharidosis and Goldenhar syndrome patients, are more likely

to experience difficult airway management. Different approaches to difficult intubation in children have been proposed, namely: fiberoptic bronchoscopy, laryngeal mask airway (LMA) and invasive procedures like retrograde intubation and tracheostomy^{7,8}.

The use of reverse LMA technique could solve the problem of difficult intubation and securing airway in KFS patients. Due to limitation of neck movements and condensed cervical vertebrae in KFS patients, reverse technique of LMA insertion as reported in the present case could be a good alternative to endotracheal intubation.

In conclusion, we suggest that reverse technique is an acceptable way of inserting LMA in neonates with Klippel-Feil syndrome. To the best of our knowledge, this is the first case reported in the literature on the use of reverse LMA technique in KFS patient.

References

1. JONES KL, SMITH S: *Recognizable patterns of human malformation*. 5th ed. Philadelphia: WB Saunders Company; pp. 320-33, 1997.
2. NAGUIB MG, MAXWELL RE, CHOU SN: Identification and management of high-risk patients with Klippel-Feil syndrome. *J Neurosurg*; 61(3):523-30, 1984.
3. SAHIN A, CEKIRGE S AND AYPAR U: Anterograde endotracheal intubation with a laryngeal mask airway and guide wire in an infant with micrognathia. *The Turkish J of Pediatrics*; 45:78-79, 2003.
4. BRAIN, AIJ: *The Intavent Laryngeal mask Instruction Manual*, 2nd edition. Henley on Themes, Intavent International SA, 1992.
5. MIZUSHIMA A, WARDALL GJ, SIMPSON DC: The laryngeal mask airway in infants. *Anaesthesia*; 47:849-851, 1992.
6. PATERSON SJ, BYRNE PJ, MOLESKY MG, SEAL RF, FINUCANE BT: Neonatal resuscitation using the Laryngeal Mask airway. *Anesthesiology*; 80:1248-1253, 1994.
7. PATEL D, MAEKIN GH: Paediatric airway management. *Current Anaesthesia Critical Care*; 11:262-268, 2000.
8. HARVEY SC, FISHMAN RL, EDWARDS SM: Retrograde intubation through a laryngeal mask airway. *Anesthesiology*; 85:1503-1504, 1996.

