

Fiberoptic intubation through laryngeal mask airway for management of difficult airway in a child with Klippel–Feil syndrome

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ABSTRACT

The ideal airway management modality in pediatric patients with syndromes like Klippel-Feil syndrome is a great challenge and is technically difficult for an anesthesiologist. Half of the patients present with the classic triad of short neck, low hairline, and fusion of cervical vertebra. Numerous associated anomalies like scoliosis or kyphosis, cleft palate, respiratory problems, deafness, genitourinary abnormalities, Sprengel's deformity (wherein the scapulae ride high on the back), synkinesia, cervical ribs, and congenital heart diseases may further add to the difficulty. Fiberoptic bronchoscopy alone can be technically difficult and patient cooperation also becomes very important, which is difficult in pediatric patients. Fiberoptic bronchoscopy with the aid of supraglottic airway devices is a viable alternative in the management of difficult airway in children. We report a case of Klippel-Feil syndrome in an 18-month-old girl posted for cleft palate surgery. Imaging of spine revealed complete fusion of the cervical vertebrae with hypoplastic C3 and C6 vertebrae and thoracic kyphosis. We successfully managed airway in this patient by fiberoptic intubation through classic laryngeal mask airway (LMA). After intubation, we used second smaller endotracheal tube (ETT) to stabilize and elongate the first ETT while removing the LMA.

Keywords: *Fiberoptic intubation, Klippel-Feil syndrome, laryngeal mask airway*

Keymessages: *The ideal airway management modality in pediatric patients with syndromes like Klippel-Feil syndrome is a great challenge and is technically difficult for an anesthesiologist. Fiberoptic bronchoscopy alone can be technically difficult and patient cooperation also becomes very important, which is difficult in pediatric patients. Fiberoptic bronchoscopy with the aid of supraglottic airway devices is a viable alternative in the management of difficult airway in children*

INTRODUCTION

Klippel-Feil Syndrome (KFS), also known as synostosis of cervical spines, is a rare (1 in 42,000) congenital anomaly of the cervical spine.^[1] Half of the patients present with the classic triad of short neck, low hairline, and fusion of the cervical spine. KFS is one among the causes of congenital syndromes with difficult airway. The ideal

airway management modality in pediatric patients with syndromes like KFS is a great challenge and is technically difficult for an anesthesiologist.^[2] Associated anomalies which include scoliosis or kyphosis, cleft palate, respiratory problems, and congenital heart diseases may further add to the difficulty.^[3]

Cleft palate may be associated with various syndromes like Pierre Robin's sequence and many other congenital anomalies.^[4] Fiberoptic bronchoscopy is ideal for endotracheal intubation in such patients as it eliminates the risk of neurological injury. Fiberoptic bronchoscopy alone can be technically difficult and patient cooperation also becomes very important, which is difficult in pediatric patients. Fiberoptic bronchoscopy with the aid of supraglottic airway devices is a viable alternative in the management of difficult airway in children.^[5]

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We report a case of KFS in an 18-month-old girl posted for cleft palate surgery.

CASE REPORT

An 18-month-old child weighing 10 kg presented with short neck and severe restriction of neck movements. Radiograph of spine revealed complete fusion of the cervical vertebrae with hypoplastic C3 and C6 vertebrae and thoracic kyphosis [Figure 1]. Computed tomography (CT) scan showed occipital meningocele. Examination of the airway showed limited flexion and extension of the upper cervical spine with adequate mouth opening. The systemic examination was normal. All other investigations were within normal limits.

Anesthetic plan was fiberoptic intubation through laryngeal mask airway (LMA), as these patients are more prone for neurological injury during intubation due to restricted neck mobility.^[6] Child was premedicated with oral midazolam (0.5 mg/kg); general anesthesia was induced with inhalation of 8% sevoflurane in 50% nitrous oxide and oxygen. Meanwhile, standard monitors of electrocardiogram, pulse oxymetry and non-invasive blood pressure were attached and intravenous access was secured with a 24-G cannula on the right hand. Manual in-line stabilization of neck was maintained by an assistant. After adequate depth of anesthesia, classic LMA size 02 was inserted and placement confirmed with bilateral air entry and square wave capnography. Inj. vecuronium 1 mg i.v. was given. A size 4 uncuffed endotracheal tube (ETT) was fitted at the upper end of the size 4 mm armored ETT and slipped over the fiberoptic bronchoscope [Figures 2 and 3].

The fiberoptic bronchoscope with the ETTs loaded over it was inserted through the LMA. The fiberoptic bronchoscope was introduced through the vocal cords and advanced into the trachea without any difficulty. Tracheal lumen was confirmed, and the armored ETT was slid from the bronchoscope and advanced into the trachea with the aid of the uncuffed ETT uneventfully, confirmed by fiberoptic bronchoscopy and capnography on ventilation. The bronchoscope, LMA, and uncuffed ETT were carefully removed in succession. Throughout the procedure of intubation, oxygen saturation of the patient was maintained. Anesthesia was maintained with sevoflurane, inj. fentanyl, and inj. vecuronium. Surgery proceeded uneventfully, and neuromuscular blockade was reversed with glycopyrrolate 0.1 mg and neostigmine 0.5 mg. The tracheal tube was removed after resumption of spontaneous breathing and return of good muscle tone. The perioperative period was uneventful with no airway-related complications.

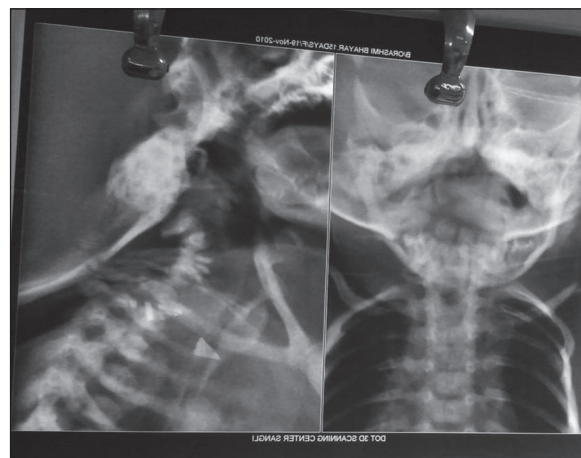


Figure 1: Radiograph of neck showing fused, hypoplastic cervical vertebra

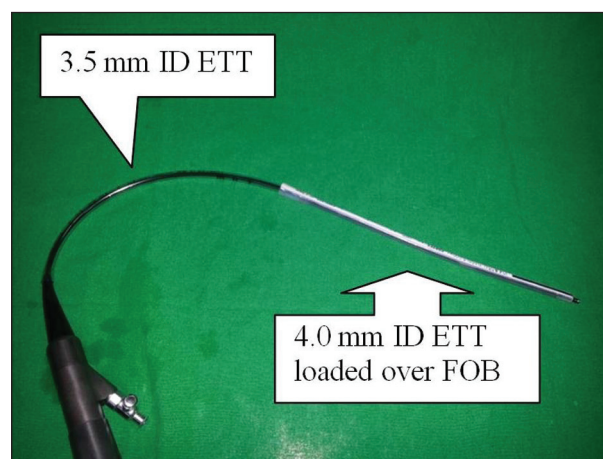


Figure 2: Two ETTs loaded over Fiberoptic bronchoscope (FOB)

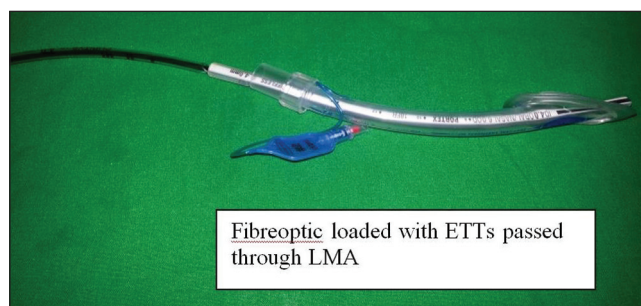


Figure 3: FOB loaded with ETT through LMA

DISCUSSION

KFS, also known as synostosis of cervical spines, is a rare congenital anomaly of the cervical spine. Short neck with limited range of movement and cervical instability can lead to neurological damage while intubation and positioning during surgery.^[6] KFS occurs as a result of failure in normal segmentation of cervical mesodermal somites

during the 2nd and 8th weeks of embryonic development, with the etiology being unknown. It most commonly presents with the triad of short neck, low hairline, and fusion of the cervical spine. Decreased neck mobility is the most common physical finding. The hypomobility between fused vertebral segments puts these patients at risk for either spontaneous neurological injury or injury due to minor trauma. Most neurological manifestations are secondary to chronic compression of cervical spinal cord, pons, medulla, and stretching of cranial nerves. Sudden neck movements or minor falls can cause basilar artery insufficiency and syncope. Hence, these patients pose a potential challenge to the anesthesiologist with regard to the management of the difficult airway. Ideally, in these patients, to avoid injury to the cervical spine, fiberoptic intubation is recommended, which ensures safe and sure airway control. Techniques which can be considered in such patients are a) fiberoptic intubation, b) retrograde-assisted fiberoptic,^[7] c) retrograde alone, d) fiberoptic intubation through LMA,^[8] e) flexible LMA alone,^[9] f) intubating LMA,^[10] and g) intubation through classic LMA.^[11] For retrograde intubation with or without assisted fiberoptic, landmarks may be vague and will be more cephalad in younger children. Thus, each technique has its own advantages and limitations. Fiberoptic intubation with the aid of LMA proves to be an ideal option in such cases, as LMA gives an additional advantage of oxygenation to the patient. Many methods can be used to take out the LMA after intubation, such as guide wire passed through the side port of fiberoptic bronchoscope,^[12] connecting another smaller size ETT to ETT inside LMA^[12] and crook airway exchange catheter.^[13] This method can be considered in other cervical spine disorders as well, e.g. achondroplasia, Morquio's syndrome, isolated odontoid anomalies, Down syndrome, spondyloepiphyseal dysplasia.

CONCLUSION

We reported here successful airway management in an 18-month-old child with fiberoptic endotracheal intubation with the aid of LMA. To conclude, fiberoptic intubation through LMA may be considered for managing difficult

airway in children with syndromes like KFS and Pierre-Robin syndrome.

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