The success of direct laryngoscopy in children with Klippel-Feil Syndrome

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Dear Editor,

Klippel-Feil Syndrome (KFS), that is characterized by severe restriction of the neck motion as a result of fusion of cervical vertebrae may impede successful airway management. Other skeletal deformities like short neck, spinal canal stenosis or scoliosis and the risk for neurological injury during positioning may complicate direct laryngoscopy (DL) and tracheal intubation.

Here, we want to report the airway management of a child with KFS (written parental consent for publication of this case has been obtained) and review the literature to investigate if there is a positive correlation between younger age and successful DL in children with KFS.

A 3 year-old girl (10 kg in weight) with KFS was scheduled for adeno-tonsillectomy. She had the classic triad of KFS with short neck, low posterior hairline and cervical fusion at C2 and C3 (Figure 1). She also had increased cervical lordosis, Sprengel deformity (elevation of the scapula due to failure of descent during fetal life), occipital encephalocele and strabismus with a Mallampati II-III.

After IV midazolam 0.5 mg she was taken to the operating theatre. Electrocardiogram, pulse oximetry and non-invasive

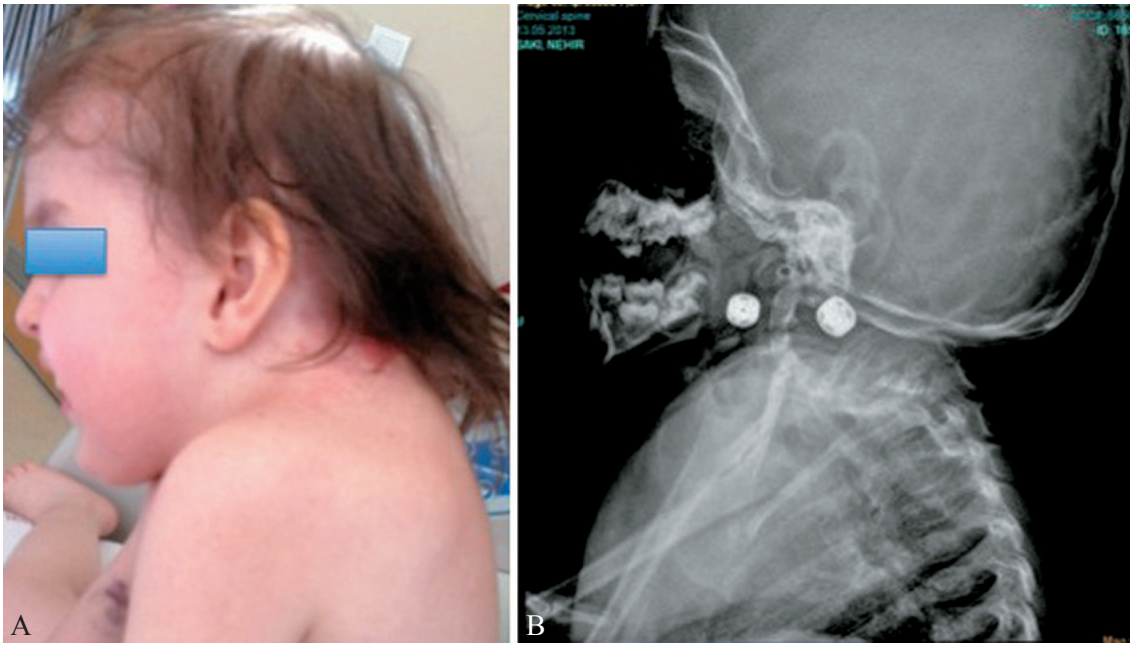

Figure 1.—A) Three-year old girl with Klippel-Feil Syndrome; B) cervical radiograph.

Table I.—*The overview of direct laryngoscopy for tracheal intubation in pediatric patients with Klippel-Feil Syndrome in literature.*

Reference number (year)	Number of patients	Age	Features	Direct laryngoscopy
1. (1974)	8	<1 year	Infants with occipital encephalocele, 6 of them also had cleft palate	Successful DL with regular laryngoscope, OTI or NTI at lateral position
2. (1986)	1	3 weeks	Occipito-cervical encephalomyelocele	Successful DL with regular laryngoscope, OTI at lateral position
3. (2001)	1	4 years*	Escobar syndrome, cleft palate, micrognati, history of unsuccessful awake DL and fiberoptic intubation when 16-month-old	Unsuccessful DL, intubation was also failed with a pediatric Trachlight™, a LMA was placed and a tracheal tube advanced through the LMA
4. (2003)	1	12 years	History of successful DL when 12 and 23 years old	Unsuccessful DL when 45 years old (multiple attempts with different blade types and sizes)
5. (2005)	1	16 years*	Cleft palate	Unsuccessful DL with regular laryngoscope, C&L grade IV, OTI with EID
6. (2006)	1	12 years	Arnold-Chiari malformation, Mallampati Score: 4	Successful DL with regular laryngoscope, OTI at first attempt (without NMB)
7. (2008)	4	10-16 years	Varied vertebral and/or chest wall anomalies	Successful DL with regular laryngoscope, OTI at first or second attempt using Macintosh blade
7. (2008)	1	8 years	Past tracheostomy	Fiberoptic intubation attempted and failed and converted to DL with regular laryngoscope, OTI at second attempt using Macintosh blade
7. (2008)	1	13 years*	Multiple craniofacial surgeries, history of difficult intubation	Unsuccessful DL (single attempt), fiberoptic intubation at 3rd attempt
8. (2010)	1	5 years	Down syndrome	Successful DL with regular laryngoscope, OTI at first attempt, C&L grade I
9. (2012)	1	6 years	History of successful OTI with DL, Mallampati Score: 4	DL was not tried. Fiberoptic intubation through LMA
10. (2012)	1	12 years*	Lingual tonsil hypertrophy	Multiple DL and fiberoptic intubation attempts, NTI with fiberoptic
11. (2012)	1	6 years	Mallampati Score: 2	Successful DL with regular laryngoscope, OTI at first attempt, C&L grade I
12. (2014)	1	7 years	Mouth opening: 4 cm, Mallampati Score: 4	Successful DL, OTI at first attempt, C&L grade III
13. (2015)	1	26 days	Modified Mallampati Score: 2	Successful DL, OTI at first attempt, C&L grade II

*unsuccessful direct laryngoscopy; DL. direct laryngoscopy; OTI: oro-tracheal intubation; NTI: naso-tracheal intubation; C&L: Cormack and Lehane; EID: endotracheal intubation device with a camera; NMB: neuromuscular blocking agent; LMA: laryngeal-mask airway.

blood pressure were applied. Fiberoptic assisted endotracheal intubation (with railroading technique) under general anesthesia while maintaining spontaneous ventilation was planned as the first approach for the airway management of the patient, but other alternative equipment and surgical team for urgent tracheotomy were also ready. After intravenous atropine 0.1 mg, anesthesia was induced with slow IV injection of lidocaine 20 mg, propofol 40 mg to ensure mask ventilation. There was no difficulty for assisted or controlled ventilation with facemask and no need for an oropharyngeal airway in its neutral position of the head and neck, so we decided to perform DL first. Anesthesia was deepened with 8% sevoflurane and fentanyl 15 μg and endotracheal intubation was performed with DL during manual stabilization of the neck. The laryngoscopy was graded as Cormack-Lehane Grade I. The position of the endotracheal tube was confirmed by capnography and auscultation.

The experience of the anesthesiologist and the type of surgery plays a key role in airway management of children with KFS. In adult patients with KFS, the safest option for securing the airway is awake fiberoptic intubation, but this can be challenging in pediatric cases. It is usually easy to manage the airway by mask or laryngeal mask airway (LMA) in children with KFS, while there is no report describing an unsuccessful or difficult mask ventilation or LMA insertion in the literature. The attempt of DL (successful or not) in pediatric patients with KFS was described in many cases in the literature [1-13] and the success of DL for tracheal intubation in these patients is overviewed in Table I. A previous uneventful anesthesia experience or successful DL does not ensure ease of securing the airway in patients with KFS because cervical fusion becomes progressively worse over time.[4, 14] However, there is no report of an unsuccessful DL in children with KFS younger than 4 years. The children who had unsuccessful DL, had accompanying conditions which were also other predictors of difficult intubation, such as Escobar syndrome,[3] cleft palate,[5] lingual tonsil hypertrophy.[10] Despite her formidable appearance for airway management, this data encouraged us to perform DL instead of fiberoptic intubation in our case.

According to the literature, the success rate of tracheal intubation with DL in patients with KFS in early ages (probably before adolescence) seems to be increased when other predictors of difficult intubation does not accompany. Nevertheless, devices for difficult airways should always be promptly available in these patients.

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