Repeated Anesthetic Management for a Patient With Klippel-Feil Syndrome

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Klippel-Feil syndrome (KFS) is a rare disease characterized by a classic triad comprising a short neck, a low posterior hairline, and restricted motion of the neck due to fused cervical vertebrae. We report repeated anesthetic management for orthognathic surgeries for a KFS patient with micrognathia. Because KFS can be associated with a number of other anomalies, we therefore performed a careful preoperative evaluation to exclude them. The patient had an extremely small mandible, significant retrognathia, and severe limitation of cervical mobility due to cervical vertebral fusion. As difficult intubation was predicted, awake nasal endotracheal intubation with a fiberoptic bronchoscope was our first choice for gaining control of the patient's airway. Moreover, the possibility of respiratory distress due to postoperative laryngeal edema was considered because of the surgeries on the mandible. In the operating room, tracheotomy equipment was always kept ready if a perioperative surgical airway control was required. Three orthognathic surgeries and their associated anesthetics were completed without a fatal outcome, although once the patient was transferred to the intensive care unit for precautionary postoperative airway management and observation. Careful preoperative examination and preparation for difficult airway management are important for KFS patients with micrognathia.

Key Words: Klippel-Feil syndrome; Micrognathia; Orthognathic surgeries; General anesthesia.

Klippel-Feil syndrome (KFS) was first described by Klippel and Feil in 1912 as a classic triad comprising a short neck, a low posterior hairline, and restricted motion of the neck due to fused cervical vertebrae.¹ The anatomical characteristics of KFS have significant implications for airway management because of the potential for difficult intubation.² In addition, patients with KFS are also more likely to have a variety of other associated anomalies.¹ We report the repeated anesthetic management for multiple staged orthognathic surgeries for a KFS patient with micrognathia.

Received June 4, 2012; accepted for publication May 20, 2014.

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Anesth Prog 61:103–106 2014

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CASE REPORT

A 27-year-old woman (160 cm, 44 kg) was scheduled for surgical treatment of her skeletal maxillary protrusion with mandibular retrognathia. She had an extremely small mandible and significant retrognathia, with an overjet (horizontal distance between the maxilla and mandible) of 18 mm. She could not make a forward motion of the mandible, and cervical extension was limited. Airway examination revealed that her Mallampati score was class 4, although she did not have a short neck. Mouth opening was 30 mm at this first visit; however, her temporomandibular joint acted only as a hinge, and it was not possible for it to glide forward. Spirometry showed slight restrictive ventilatory impairment resulting from thoracic hypoplasia and a vital

ISSN 0003-3006/14
SSDI 0003-3006(14)

capacity of 73%. Her vital signs, electrocardiogram, and blood tests were within normal limits. Other physical findings included hypoplasia and asymmetry of the clavicle and scapula, with limited ability to raise her arms.

Orthopedic examination revealed severe limitation of cervical mobility, with only 20° of head flexion and extension and 20–30° of lateral rotation possible, due to fusion of the C3–5 vertebrae. However, according to an orthopedician's evaluation, the normal range of neck motion required for endotracheal intubation during anesthetic induction was unlikely to cause any neurological damage, as her X-ray showed no basilar invagination or cervical cord compression. Neurosurgical examination demonstrated atlantoaxial subluxation, but with no spinal compression or cervical unsteadiness. In addition, her magnetic resonance imaging and echoencephalograph revealed no cerebral abnormalities, and she had no neurological symptoms. She did not complain of either spontaneous pain or pain on motion in the back of the head and neck. Urological examination revealed no renal dysfunction. Otolaryngological examination showed that tracheotomy would be possible normally, and a deviated nasal septum and mild narrowing of the pharyngeal cavity were noted. There was no other significant positive medical or surgical history.

We considered a multiple-contingency plan for induction of anesthesia and airway management. In view of the severity of her micrognathia and cervical spine deformity, we planned an awake fiberoptic intubation. Written informed consent for the procedure was obtained from the patient, after explaining the risks involved in securing her airway. She was eager to have the orthognathic surgery even if tracheotomy was unavoidable in the perioperative period. Staged surgeries were planned because of the large displacement of the gnathic bones. Before each surgery, an intensive care unit bed was reserved for her in the eventuality of respiratory failure due to upper airway obstruction in the postoperative period. As a precaution, tracheotomy equipment was always kept ready in the operation room. Four days before the operation, her nasal cavity and pharynx were examined with a fiberoptic bronchoscope. As a result, anatomical factors causing difficulty for nasal intubation using a fiberoptic bronchoscope were not visualized, and thus we expected that the probability of successful intubation was high.

Surgically assisted rapid palatal expansion was planned as the initial surgery. The patient's airway was topically anesthetized with 2% viscous lidocaine (mouth-wash and gargle), 4% lidocaine solution (a percutaneous injection into the pharynx), and 8% topical lidocaine solution sprayed directly onto the mucosa of mouth, tongue, and nasopharynx and then left for 5–15 minutes in the region of mucosa until these anesthetics took

effect completely. Also, 0.2% phenylephrine as a vasoconstrictor and 2% lidocaine gel was used in the nasal cavity to help prevent nasal bleeding and to provide topical anesthesia in the nose.

Awake nasal intubation (size 6.5 mm cuffed Portex Ivory tube) using a fiberoptic bronchoscope was smoothly completed in the first attempt under moderate sedation where the patient was still able to take a deep breath when asked. Using fentanyl (50 µg) and propofol (effect-site target concentration was 1.5–2.0 µg/mL using a target-controlled infusion [TCI] pump), we expected that we could intubate safely under moderate sedation. After intubation, general anesthesia was induced with fentanyl (50 µg), propofol (121 mg), and rocuronium (50 mg). Anesthesia was maintained with air, oxygen, and sevoflurane (fraction of inspired oxygen [FiO₂] = 0.4) under controlled ventilation. Direct laryngoscopy with a Macintosh laryngoscope was attempted for the purpose of airway assessment after induction, and it revealed just the soft palate, ie, Cormack and Lehane grade 4, defined as that neither the glottis nor the epiglottis could be seen. The surgery lasted for 1 hour 47 minutes, at the end of which the patient was extubated with no difficulty when she was completely awake. After extubation, she did not suffer respiratory failure and had an uneventful postoperative course.

The second surgery, mandibular distraction osteogenesis, was planned 2 years after the first procedure. Fiberoptic bronchoscopy-assisted awake nasal intubation was successfully performed, as with the first surgery. Anesthesia was maintained with total intravenous anesthesia (TIVA) using propofol (TCI 3–4 µg/mL; total amount was 3500 mg) and remifentanyl (0.1–0.3 µg/kg/min, 7.9 mg total dose) to avoid increased risk of vomiting compared to the first procedure and because the scheduled surgical time was 5 hours. Actually, the surgery was extended to 12 hours, and the patient was transferred to the intensive care unit with the endotracheal tube left in situ. In the intensive care unit, artificial ventilation was performed under propofol sedation. There was still the risk of airway obstruction because of the limited mouth opening and the possibility of laryngeal edema, though the patient could open her mouth to some extent because of the presence of surgical internal fixation. Because there was no worsening of the swelling noted by fiberoptic bronchoscopy, a cuff-leak test was performed where the patient was able to breathe around the purposefully occluded endotracheal tube after the cuff was deflated. Because it was also confirmed that there were no residual effects of narcotics or muscle relaxants, she was extubated after several hours and was transferred to a general dental ward the next day without any complications.

Four months after the second surgery, removal of the distractor and fixed plate was planned. This time, because of an infection around the right side of the distractor, the patient's mouth opening was restricted to 10 mm. We were again able to accomplish awake nasal intubation using the fiberoptic bronchoscope without difficulty. Direct laryngoscopy after induction revealed Cormack and Lehane grade 4 even after the surgery of mandibular distraction. Anesthesia was maintained with TIVA to reduce the risk of vomiting during the operation, which ended after about 5 hours. After we confirmed the absence of airway edema by using the fiberoptic bronchoscope, the patient was extubated when she met all extubation criteria.^{3,4} Although we considered that the risk of postoperative respiratory failure would be somewhat less than previously, and that it would be possible to reintubate her by fiberoptic bronchoscope if needed in an emergency, we nevertheless waited until she was completely awake before she was extubated. She was later transferred to the general dental ward without any difficulty.

DISCUSSION

KFS patients are assigned to 1 of 3 types: type 1, those with extensive fusions of many cervical vertebrae; type 2, those with fusions at only 1 or 2 cervical interspaces; and type 3, those with fusions in the cervical spine accompanied by fusion in the lower lumbar spine. Patients with type 2, which is the most common form of the disease including our patient, most often have an autosomal dominant inheritance pattern.^{1,5}

KFS can be associated with a number of other anomalies, including renal dysfunction (64%), scoliosis (60%), deafness (30%), Sprengel's scapula deformity (25–35%), congenital heart disease (4.2–14%), mental deficiency, pulmonary disability, and cleft lip and palate.^{6–10} KFS is a malformation sequence that starts as early as the fourth or beginning of the fifth week of fetal life.¹¹ KFS encompasses an increasing constellation of clinical features that affects most organs.⁶ Embryonic development of the mandible (approximately 10 weeks of life), palate (5–12 weeks), and cervical vertebrae (8 weeks) are known to occur at an approximately similar gestational age.⁸ The embryological sequence of events leading to the abnormalities of KFS appears to be failure of or abnormal fusion of the cervical vertebrae and palatal shelf, causing cleft palate or retrognathia, and the possibility of other associated anomalies around the oral area.⁶ Patients with KFS are also more likely to have cervical instability, increasing their risk of neurologic damage with minor trauma or during laryngoscopy, intubation, and positioning.⁹ Mandibular hypoplasia with

KFS can cause upper airway obstruction resulting in obstructive sleep apnea.¹² Teoh et al¹³ described associated craniofacial anomalies, such as mandibular malformations and micrognathia, may make oxygenation and ventilation difficult. Hence, successful anesthetic management of these patients requires an understanding of the anatomical and pathological changes associated with these congenital anomalies. Therefore, we performed a careful preoperative evaluation to exclude them. In this case, preoperative screening examinations were conducted in the department of urology, internal medicine, otology, and neurosurgery. These examinations excluded associated congenital anomalies of the heart and kidneys, although mild restrictive ventilatory impairment derived from thoracic deformities was revealed, which was not of sufficient severity to interfere with the anesthetic management of this patient. Further, it was determined that neck movement within the normal range of motion would not be neurologically detrimental although atlantoaxial subluxation was revealed preoperatively.

Patients with KFS pose numerous problems for anesthesiologists, one of which is difficult airway. In a previous case report of surgery in a KFS patient, the patient ultimately required emergency tracheostomy postoperatively for complete upper airway obstruction, although a blind oral intubation was successful after several attempts.¹⁴ In another case, several attempts at endotracheal intubation were associated with complications such as laryngospasm, hypoxia, and bradycardia, which required cardiac massage, and even abandonment of the surgical procedure because of unsuccessful endotracheal intubation.¹⁵ Our KFS patient had severe micrognathia in addition to limitation of neck mobility due to cervical vertebral fusion. Although her mouth opening was about 30 mm at first with only a hinge action of mandibular movement at the temporomandibular joint, it was restricted to 10 mm prior to the third surgery. Awake nasal intubation with fiberoptic bronchoscopy was our first choice for gaining control of the patient's airway because difficult intubation was predicted, and perioperative examination of the occlusal relationship was required, which ruled out oral intubation. Although we thought that the possibility of success of intubation using fiberoptic bronchoscopy was high, we prepared for a tracheotomy each time, and in addition, we had the other equipment for alternative airway management such as a laryngeal mask and percutaneous tracheal puncture needle. Direct laryngoscopy performed at each of the 3-time surgeries revealed her laryngoscopic view was Cormack and Lehane grade 4. As previously reported, an uneventful previous surgical experience does not ensure ease of securing the airway in patients with KFS, because cervical fusion becomes progressively

worse over time.¹⁶ Careful evaluations are required each time the patient presents for surgery.

After the second surgery, the patient was transferred to the intensive care unit. Mandibular surgery has the potential to alter the skeletal structure of the airway¹⁷ and may especially increase risk of airway obstruction due to laryngeal edema postoperatively. Actually, this patient had an increased risk of soft-tissue swelling due to prolonged surgery, in addition to other risk factors for airway obstruction such as the extreme micrognathia and limitation of neck movement and mouth opening due to the hinge-only action of the mandible. Because there was an increased risk of difficult reintubation and postoperative airway obstruction that might have required emergency tracheostomy, she was followed up in the intensive care unit. She corresponded to the category of “at risk” in the Difficult Airway Society’s extubation guideline,⁴ where the threat of airway compromise is so severe that extubation should not take place immediately after surgery. Thus, it was reasonable to postpone her extubation until she had recovered sufficiently in the intensive care unit according to the extubation algorithm. Postponing extubation for a few hours, or in some cases for a few days, may be the most appropriate course of action.⁴

In conclusion, careful preoperative examination and preparation for difficult airway management are important for KFS patients with micrognathia. We also recommend elective postoperative intensive care monitoring after extensive surgery, as these patients are at a high risk of developing postoperative airway obstruction and respiratory failure.

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