

Klippel-Feil syndrome for scoliosis surgery: management of a potentially difficult paediatric airway, and report of false-negative motor-evoked potential

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Abstract

A six-year-old girl with Klippel-Feil syndrome and thoracolumbar scoliosis was scheduled for growing rod insertion. Inhalational induction and tracheal intubation were carried out, with her neck in a neutral position. However, the patient woke up with paraplegia, despite normal intraoperative neurophysiological monitoring, which necessitated immediate revision surgery. Intravenous induction was performed for the second surgery. We discuss the management of a potentially difficult paediatric airway, and report on false-negative motor-evoked potential.

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Introduction

Klippel-Feil syndrome (KFS) was first described by Maurice Klippel and André Feil in 1912 as a triad of short neck, restricted motion of neck, and low posterior hairline.¹ As a consequence of failure in normal segmentation of cervical vertebrae during the early weeks of foetal development, two or more of the seven vertebrae are fused. It has a rare incidence of 1:42 000 births, of which 65% are female. KFS is frequently associated with other anomalies, the spectrum of which is immense, ranging from congenital scoliosis (more than 50%), Sprengel's deformity (20-30%), and deafness (30%), to cardiovascular (4-29%), and genitourinary (25-35%) abnormalities. KFS becomes a challenge to anaesthesiologists when patients present for surgeries such as scoliosis correction. Severe restriction of cervical motion from fusion and cervical instability renders intubation difficult, particularly in the paediatric population, where awake fibre-optic intubation (FOI) is not an option.

Currently, intraoperative neurophysiological monitoring (IOM), for example, somatosensory-evoked potential (SSEP) and motor-evoked potential (MEP), has become a mainstay in avoiding potential neurological complications. Although the results of false negatives are very rare, potential signal depression from various anaesthetic agents could limit the reliability of these parameters. We describe the perioperative management of such a patient in a state general hospital, with an emphasis on difficult airway

management, and the rare occurrence of false-negative MEP, resulting in postoperative paraplegia.

Case report

A six-year-old, 18-kg girl with thoracolumbar scoliosis was scheduled for growing rod insertion via posterior approach. She had KFS, associated with multiple congenital anomalies: fused C6 and C7 vertebrae, occipital encephalocoele that was resected at one-year old, cervical syringomyelia, dextrocardia, atrial septal defect, horse shoe kidney, and right hand polydactyly. There was no report of weakness, or altered sensation, of both the upper and lower limbs. She also had a hypoplastic right lung, and had been admitted on multiple occasions to hospital for bronchopneumonia, as well as acute exacerbation of bronchial asthma.

The patient's systemic examination was unremarkable, except for the severity of scoliosis, and reduced right breath sound. Her airway examination showed that she had a webbed neck, and congenital muscular torticollis on the right, with very limited extension (10 degree), flexion (25 degrees) and rotation (10 degrees). However, her mouth opening was adequate. Mallampati score was two. Blood investigations were within normal limits, with haemoglobin of 12.4 g/dl, urea of 1.5 mmol/l and creatinine of 40 µmol/l. Arterial blood gas analysis was normal. Due to the unavailability of a paediatric respiratory service in the hospital, we did not subject the patient to a lung function

test. A chest radiograph revealed a hypoplastic right lung. Echocardiography confirmed dextroposition of the heart, a small secundum atrial septal defect (3 mm), and absence of the left pulmonary artery. Magnetic resonance imaging reported severe thoracolumbar scoliosis, and fused C6 and C7 vertebrae, with no significant spinal canal narrowing. Preoperative Cobb's angles were 60 degrees (T5-T10) and 45 degrees (T10-L3).

Written parental anaesthetic consent was obtained, with an explanation that focused on the potentially difficult airway, neurological injury, and prolonged postoperative ventilation, ascribed to the distorted anatomy and limited respiratory reserve. The patient was nebulised with salbutamol as premedication. Prior to surgery, she was nebulised with 1 ml of lignocaine in 2 ml normal saline, and given intravenous (IV) glycopyrrolate 5 µg/kg as a preparation for asleep FOI. An otorhinolaryngology (ORL) surgeon was on standby in the operating room to carry out a tracheostomy in the event of failed airway control.

Inhalational induction was performed, with an incremental dose of sevoflurane up to a concentration of 5% in 100% oxygen at 6 l/minute, under American Society of Anesthesiologists' standard monitoring. Mask ventilation was easy, and spontaneous ventilation maintained. After adequate depth of anaesthesia was achieved, IV fentanyl 2 µg/kg and IV lignocaine 1 mg/kg were given. Direct laryngoscopy was attempted with a second anaesthesiologist providing manual in-line stabilisation of the neck. Further topicalisation with lignocaine 10% (two sprays) was given to obtund sympathetic response. A Cormack and Lehane Grade 1 view of the glottic inlet was observed, and tracheal intubation was carried out without difficulty.

IV vecuronium 0.1 mg/kg was administered to ease positioning, as due to its cost, it was the most commonly used muscle relaxant in this centre.

Anaesthesia was maintained with sevoflurane (minimum alveolar concentration 1.0) in a mixture of 35% oxygen and air at 2 l/minute, guided by bispectral index (Aspect Medical System) kept between 40-60. IV morphine 2 mg bolus, and IV fentanyl infusion at 1 µg/kg/hour, provided the analgesia. No further muscle relaxant was given. Two large-bore intravenous accesses were secured. Cannulation of radial artery allowed continuous invasive blood pressure monitoring. The patient was carefully positioned prone, with the pressure points well protected.

Temperature was maintained with a forced-air warming blanket Bair-Hugger® and Barkey® autoline. Intraoperative multimodality neurophysiological monitoring (TECA Synergy

N-EP, Viasys Healthcare, USA) using SSEP and MEP was utilised. Any decrement of more than 50% of the baseline values, or a 10% increment in latency, was considered to be a significant change. Haemodynamic was stable, and the patient was normothermic throughout the three-hour surgery. Both SSEP and MEP also remained within normal limits. The patient was given Hartman's solution as intraoperative fluid replacement. Blood loss was estimated to be 150 ml, hence no blood transfusion was given. Postoperative haemoglobin was 11.9 g/dl.

However, the patient woke up with lower limb paralysis and paraesthesia, despite the fact that SSEP and MEP did not show any signs of cord injury. Postoperative Cobb's angles were 23 degrees (T5-T10) and 24 degrees (T10-L3). IV methylprednisolone was initiated at 30 mg over 15 minutes, followed by 5 mg/kg/hour, and immediate revision surgery was planned. Intravenous induction was performed with IV fentanyl 1 µg/kg, and propofol 1.5 mg/kg. Tracheal intubation was facilitated with vecuronium 0.1 mg/kg. Both SSEP and MEP signals of the lower limbs were absent, which returned upon release of domino screws. Cobb's angles after revision surgery were 40 degrees (T5-T10) and 34 degrees (T10-L3). The patient had full neurological recovery on day three post-surgery.

Discussion

Patients with KFS are classified into one of the three types of cervical fusion abnormalities: Type I, those with extensive fusion of many cervical vertebrae; Type II, those with fusions at only one, or two, cervical interspaces; and Type III, those with fusions in the cervical spine, accompanied by fusions in the lower lumbar spine.¹ Three patterns of cervical fusions are associated with higher risk of instability and injury: C2-C3 fusion with occipitalisation of the atlas; long fusion and abnormal occipitocervical junction; and a single open space between two fused segments.

Our patient only had fused C6 and C7 vertebrae, but her congenital muscular torticollis and short neck further contributed to her restricted cervical motion. Any major movement during airway manipulation and operative positioning could result in neurological sequelae.^{2,3} Her limited respiratory reserve from the hypoplastic right lung and hyper-reactive airway disease further complicated the matter. Any delay in securing a tracheal tube could potentially cause rapid deterioration of oxygen saturation.

Even though this patient underwent uneventful general anaesthesia at the age of one, it did not ensure the ease of securing the airway, as her cervical fusion may progressively worsen over time.³ We chose inhalational induction with sevoflurane, as it maintains spontaneous

respiration. It also has a relatively fast onset and offset. We attempted manual ventilation, and it was successful. We then proceeded to direct laryngoscopy with manual in-line stabilisation, once adequate depth of anaesthesia was achieved. Our subsequent plan would have been asleep FOI if direct laryngoscopy appeared to be difficult, thus her airway was topicalised with nebulised lignocaine prior to induction, and IV glycopyrrolate was given to reduce airway secretions. Asleep FOI under inhalational anaesthesia, with maintenance of spontaneous ventilation, may be chosen as the first modality, if not as an alternative option. An ORL surgeon was also available to provide surgical airway access in the event of failed intubation, or mask ventilation. The multiple contingency plans for induction of anaesthesia and airway control are depicted in Figure 1. Delivery of the second anaesthesia was more straightforward, with conventional IV induction, in view of an uneventful prior anaesthesia. Anaesthesia was maintained as per the first surgery.

There is a case series of 10 paediatric patients with KFS, who had undergone general anaesthesia, and who were easily managed with conventional approaches, such

as direct laryngoscopy followed by tracheal intubation, laryngeal mask airway placement, or mask ventilation.⁴ Only one patient required FOI that was difficult. These cases involved paediatric patients, who may have more cervical flexibility, in comparison to adult patients. Nevertheless, a comprehensive preoperative airway assessment to identify potential difficulty and risk of neurologic injury, and development of a primary airway management plan and its major contingency backups, are key with regard to management, as illustrated in our case study.

IOM can provide real-time data on neurologic status to guide intraoperative manoeuvres and decisions. Traditionally, SSEP has been the mainstay of IOM. However, SSEP only directly monitors the ascending (sensory) pathways within the spinal cord. It may fail to reflect spinal cord injury that affects the descending (motor) pathways. Several case reports have showed the presence of preserved SSEP with postoperative paraplegia.⁵ MEP, which monitors abnormality in motor pathways, is more sensitive to spinal cord ischaemia, and has several advantages when compared to SSEP.⁶ Despite the literature showing the high reliability of MEP,⁷ there are sporadic case reports, though

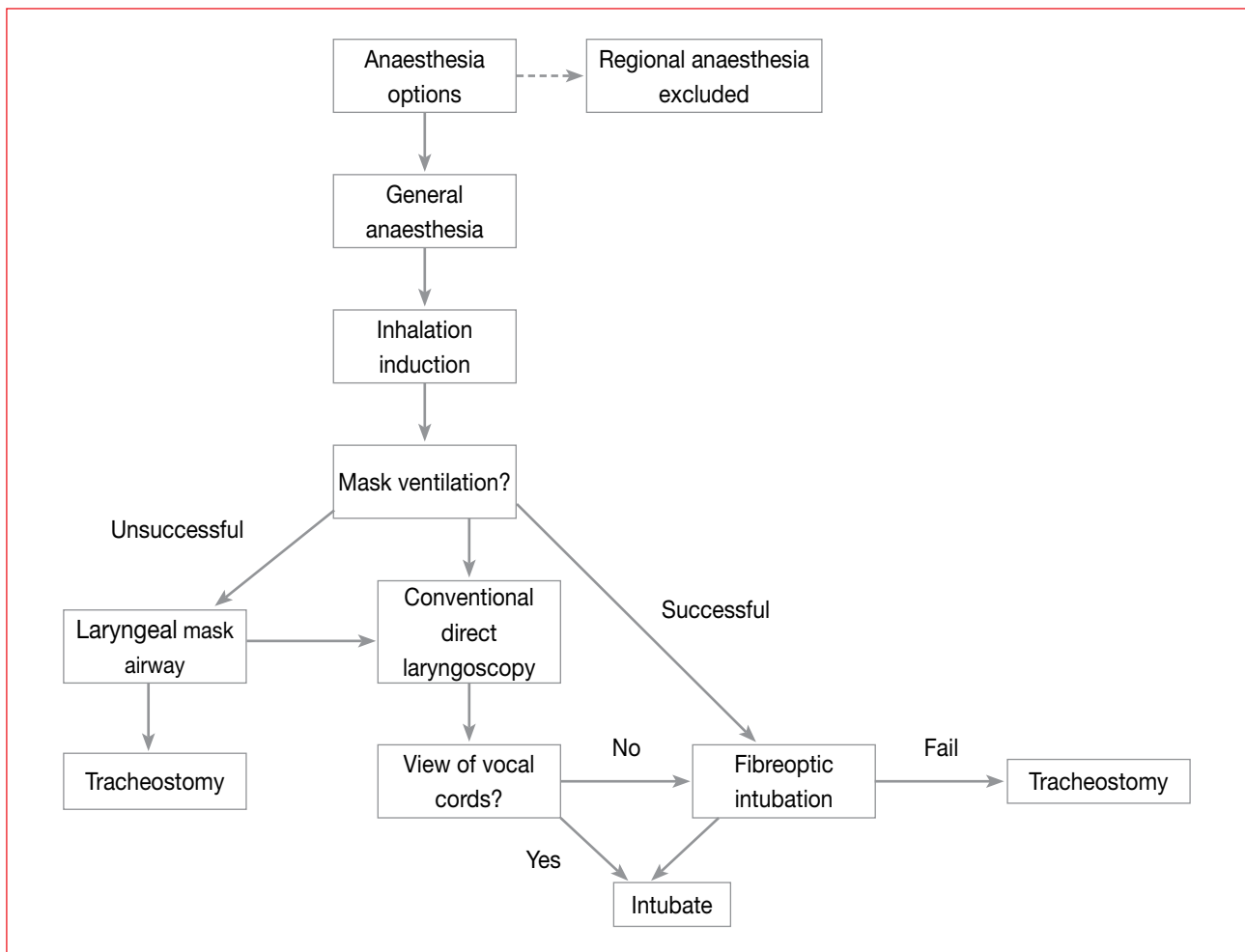


Figure 1: Flowchart showing the options considered for management of the airway

rare, about false-negative MEP during scoliosis surgery, causing paralysis.^{8,9}

Combined SSEP and MEP monitoring was reported by some authors to be reliable, with sensitivity of 98.6%, and specificity of 100%.¹⁰ In fact, multimodal IOM has proven to allow more accurate detection of neurologic status than the single modality technique that has become the standard of care in spine surgery.¹¹ In our case, both SSEP and MEP did not reveal any signs of cord injury throughout the procedure. There were only delayed true-positive SSEP and MEP during the revision surgery, which recovered upon releasing of the rods in anticipation of overstretching. IV anaesthetics are the recommended maintenance anaesthetics to optimise evoked potential monitoring, as volatile agents are known to be potent depressors of the SSEP and MEP signals.¹² In our centre, we do not have target-controlled infusion pumps that are suitable for paediatric use. Though we could have used the conventional dosage calculation of propofol infusion, we opted for sevoflurane, with a target minimum alveolar concentration value not more than 1.0 intraoperatively, with bispectral index monitoring instead. Fentanyl infusion was preferred over remifentanyl, due to its cost. Blood pressure, temperature, as well as intraoperative anaemia, had been attributed to spinal cord ischaemia, but these were stable throughout the surgery, and the possibility of their implications in our case study was low.

Besides modifying anaesthetic techniques to optimise neurophysiological monitoring, anaesthesiologists should be aware of the possibility of unreliable MEP and its limitations, which may result in morbidity in patients. IOM does not eliminate all adverse neurologic events, even in the most ideal situations. Early detection of paralysis in the post-anaesthesia care unit was crucial, and immediate intervention to correct its aetiology led to successful recovery of the patient. A close working relationship among neurophysiologist, surgeon, and anaesthesiologist, is extremely important in order to deliver and interpret IOM successfully.

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