Anesthetic Management of a Patient with Klippel-Feil Syndrome for Laparoscopic Pelvic Surgery: A Case Report

Criselle C. Chua, MD and Mark Andrew B. Cruz, MD

Department of Anesthesiology, Philippine General Hospital, University of the Philippines Manila

ABSTRACT

Klippel-Feil Syndrome (KFS) continues to pose significant challenges for anesthesiologists. Beyond the expected complexities of managing difficult airways in these patients, they often present with systemic anomalies that can elevate the risk of morbidity during surgeries conducted under anesthesia. Furthermore, laparoscopic procedures bring about additional physiologic changes that must be taken into consideration when planning the anesthetic care for these individuals.

This report details the anesthetic management of a 29-year-old female diagnosed with Klippel-Feil Syndrome (KFS) and concomitant Müllerian duct aplasia-Renal agenesis-Cervicothoracic Somite dysplasia (MURCS) as well as Chiari Type 1 Malformation, who underwent a successful pelvic laparoscopic surgery. The airway was secured through awake fiberoptic-guided intubation while general anesthesia was maintained with a combination of sevoflurane inhalation and remifentanil infusion. Intraoperatively, the team prioritized neuroprotection, lung-protective ventilation strategies, and renal preservation measures.

The anesthetic management of patients with KFS necessitates a comprehensive assessment of their anomalies. Incorporating these considerations into the anesthetic management will help mitigate the procedure's adverse effects and lead to favorable patient outcomes.

Keywords: anesthesia, airway management, Klippel-Feil Syndrome, laparoscopy

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Corresponding author: Criselle C. Chua, MD
Department of Anesthesiology
Philippine General Hospital
University of the Philippines Manila
Taft Avenue, Ermita, Manila 1000, Philippines
Email: ccchua6@up.edu.ph
ORCiD: https://orcid.org/0009-0009-3355-3342

INTRODUCTION

Klippel-Feil Syndrome (KFS) is a rare inherited autosomal dominant disorder that occurs in 1 in 40,000 to 42,000 births, with an incidence of 0.71%, and a female preference.[1] Its defining characteristics include the congenital fusion of two or more cervical vertebrae, and it is clinically recognized by the triad of a short neck, a low posterior hairline, and limited neck mobility.[2,3] Additionally, individuals with KFS often present with multiple associated systemic anomalies, making it a polysyndromic condition.[4] Notably, genitourinary abnormalities affect approximately 64% of patients, significant scoliosis is observed in 60% of cases, and Chiari I malformation frequently co-occurs alongside bony abnormalities.[5,6]

We present a case of a 29-year-old female with KFS, with coexistent Müllerian duct aplasia-Renal agenesis-Cervicothoracic Somite dysplasia (MURCS) association, and Chiari Type 1 Malformation for laparoscopic excision of non-communicating rudimentary horn under general anesthesia. The difficulties encountered in the case extend beyond the

anticipated difficult airway. The anesthetic management was further complicated by potential issues arising from Chiari malformation, restrictive lung disease secondary to scoliosis, and renal agenesis associated with MURCS.

Currently, there is a paucity of literature in the anesthetic management of KFS. Specifically, there have been fewer than 50 studies published on this subject between 1974 and 2023. Remarkably, only one of these studies has explored the anesthetic management in the context of laparoscopic surgery for KFS patients.[7] No established anesthetic management guidelines currently exist for this patient population. The anesthetic plan for this particular patient was formulated through a meticulous assessment of the potential interplay between anesthesia, surgery, and her underlying syndrome.

CASE PRESENTATION

A 29-year-old nulligravid woman with a short neck and progressive skeletal deformities of the shoulder, sought consult for fertility assessment. A transvaginal ultrasound examination revealed the presence of a left endometriotic cyst, a unicornuate uterus with a non-communicating left rudimentary horn, and left renal agenesis. Subsequently, she was referred to the Genetics service, where she was diagnosed with Klippel-Feil Syndrome (KFS) in conjunction with a MURCS association. An MRI evaluation conducted to assess cervical synostosis incidentally revealed the presence of a Chiari Type I malformation. The planned surgical procedure entailed the excision of the left non-communicating rudimentary horn through operative laparoscopy. Subsequently, the patient was referred to the Anesthesia service for a preoperative evaluation.

Other than the syndrome, she had an unremarkable medical and surgical history. Examination of the airway showed a mouth opening greater than 4 cm, and a Mallampati score of 4, with no dentures nor loose teeth (Figure 1). She has a short, webbed neck, with right shoulder higher than the left, and a rib hump on the left. Thoracic levoscoliosis and lumbar dextroscoliosis were also appreciated. The rest of the systemic physical examination was unremarkable. Neurological examination showed limited cervical range of motion and shoulder shrug. There were no motor weaknesses nor sensory deficits appreciated.

MRI of the spine showed multiple vertebral fusion anomalies at C2-C7 and T1-T4 with midline fusion defects (Figure 2). Other findings included a Chiari I malformation (Figure 3), evidenced by the inferior displacement of medulla and cerebellar tonsils; platybasia, an abnormal flattening of the skull base; syringohydromyelia, fluid-filled cysts within the spinal cord; and cervical and thoracolumbar dextroscoliosis and thoracic levoscoliosis (Figure 4). MRI of the abdomen showed an absent left kidney (Figure 5).

Assessment at this time was American Society of Anesthesiologist Physical Status (ASA- PS) 2 for congenital anomalies. She had a 6% risk for major cardiac event

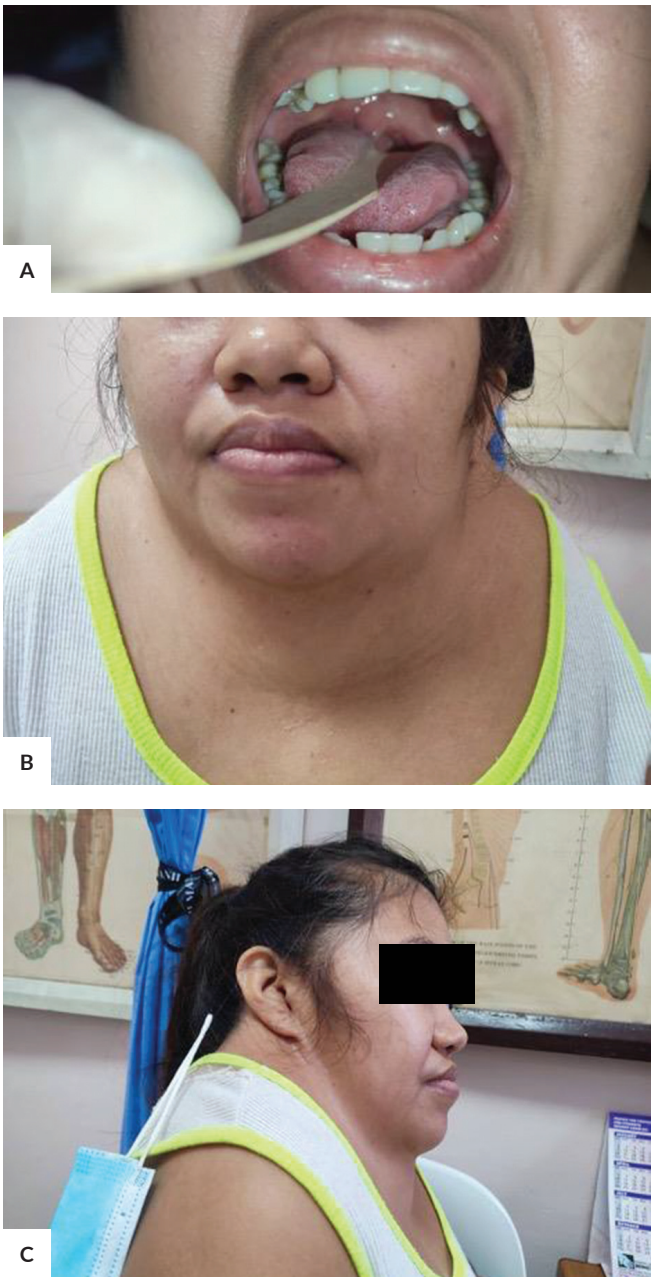

Figure 1. Airway assessment of the patient showing **(A)** Mallampati score of 4; and short, webbed neck with limited range of motion at anterior **(B)** and lateral **(C)** views.

[Class II Revised Cardiac Risk Index (RCRI)], low risk for postoperative pulmonary complications [15 points in the Assess Respiratory Risk in Surgical Patients in Catalonia (ARISCAT) Risk Index], and low risk for acute kidney injury [Simple Postoperative Acute Kidney Injury Risk (SPARK) Class A]. She had a high risk for difficult airway with an El Ganzouri Risk Index score of 6.

Following a multidisciplinary consultation, the decision was made to proceed with the surgical procedure under

general anesthesia. Given the potential risks associated with intubation and ventilation, it was determined that awake fiberoptic intubation would be the preferred approach, with the Otorhinolaryngology service on standby to perform tracheostomy if deemed necessary.

The patient was received in the operating room awake, alert and not in cardiorespiratory distress. Standard ASA monitors were attached. Plain lactated Ringer's solution was maintained at 100 ml/hr (1.8 ml/kg/hr) as the intraoperative intravenous fluid. A nasal cannula was used to deliver oxygen at 4 L/min. The head of the bed was elevated at 30 degrees. Remifentanil target-controlled infusion (TCI) intravenously was started at 0.1 ng/ml. Topical lidocaine 10% spray was applied liberally on the posterior oral cavity and posterior pharyngeal walls. An oral mouth guard was placed. An adult flexible fiberoptic scope (Karl Storz Intubation Fiberscope),

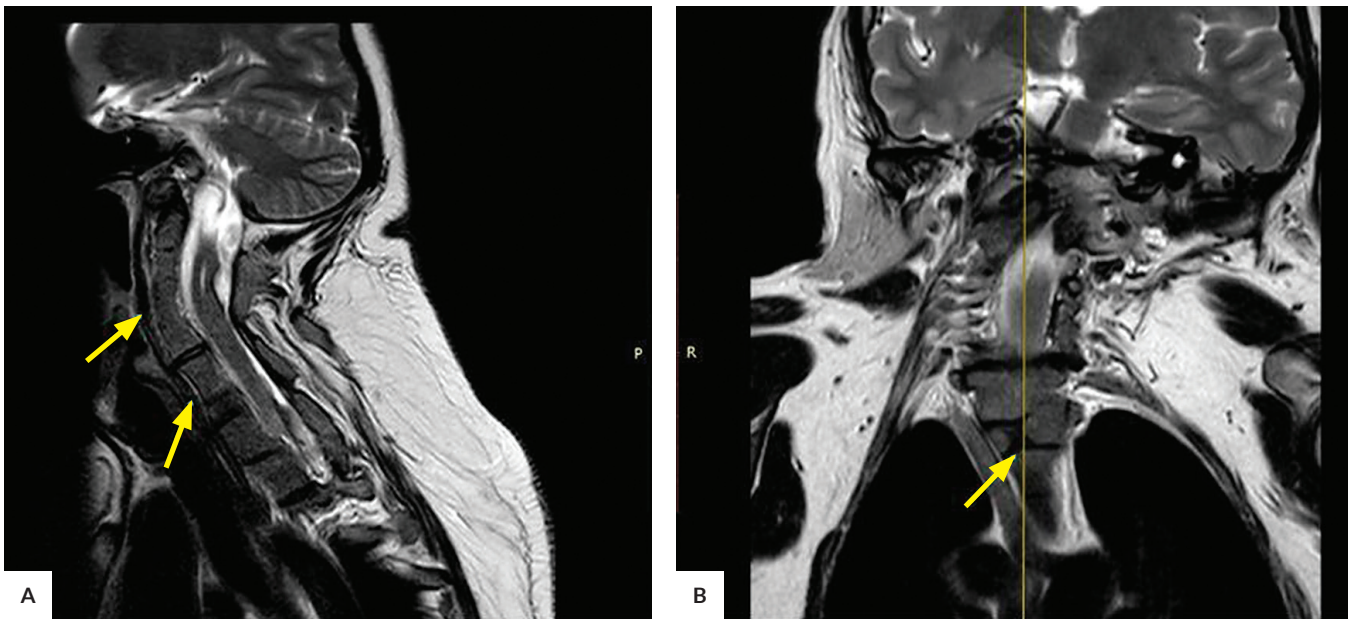

Figure 2. MRI of the spine showing **(A)** multiple vertebral fusion anomalies at C2- C7 *(yellow arrows)* and **(B)** midline fusion defects *(yellow arrow)* compatible with Klippel-Feil Syndrome.

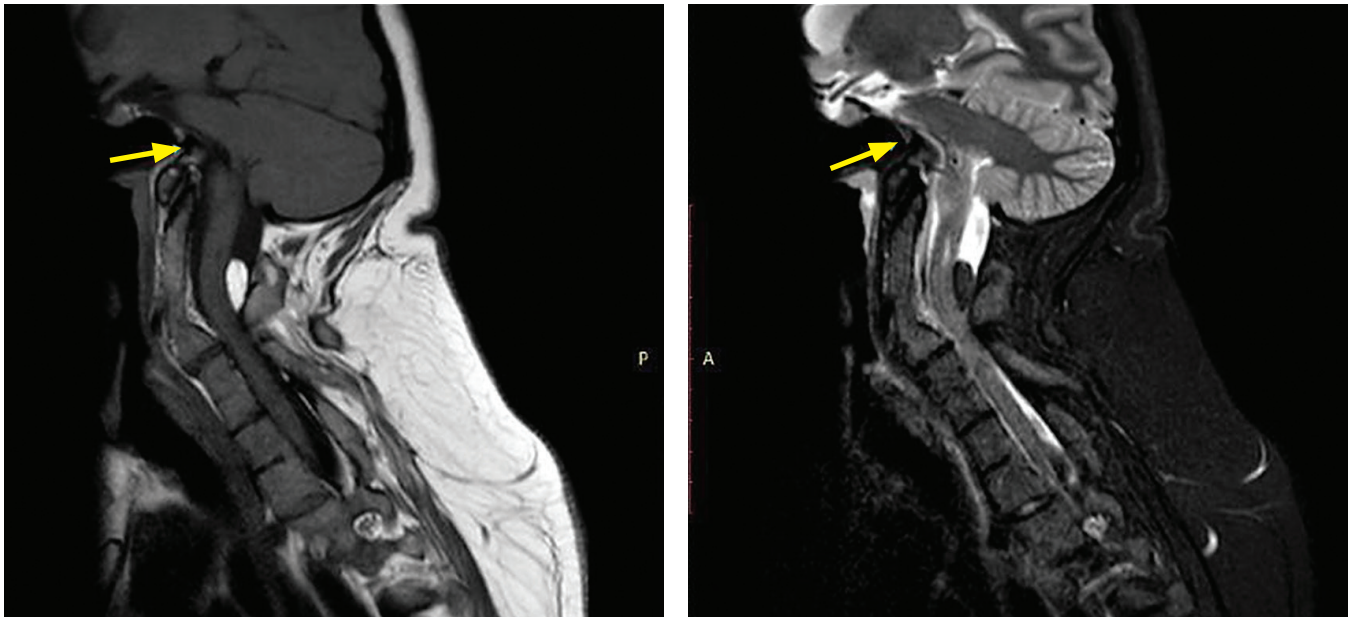

Figure 3. MRI of the spine showing Chiari Type 1 malformation as evidenced by the inferior displacement of medulla and cerebellar tonsils *(yellow arrows)*, platybasia, and syringohydromyelia.

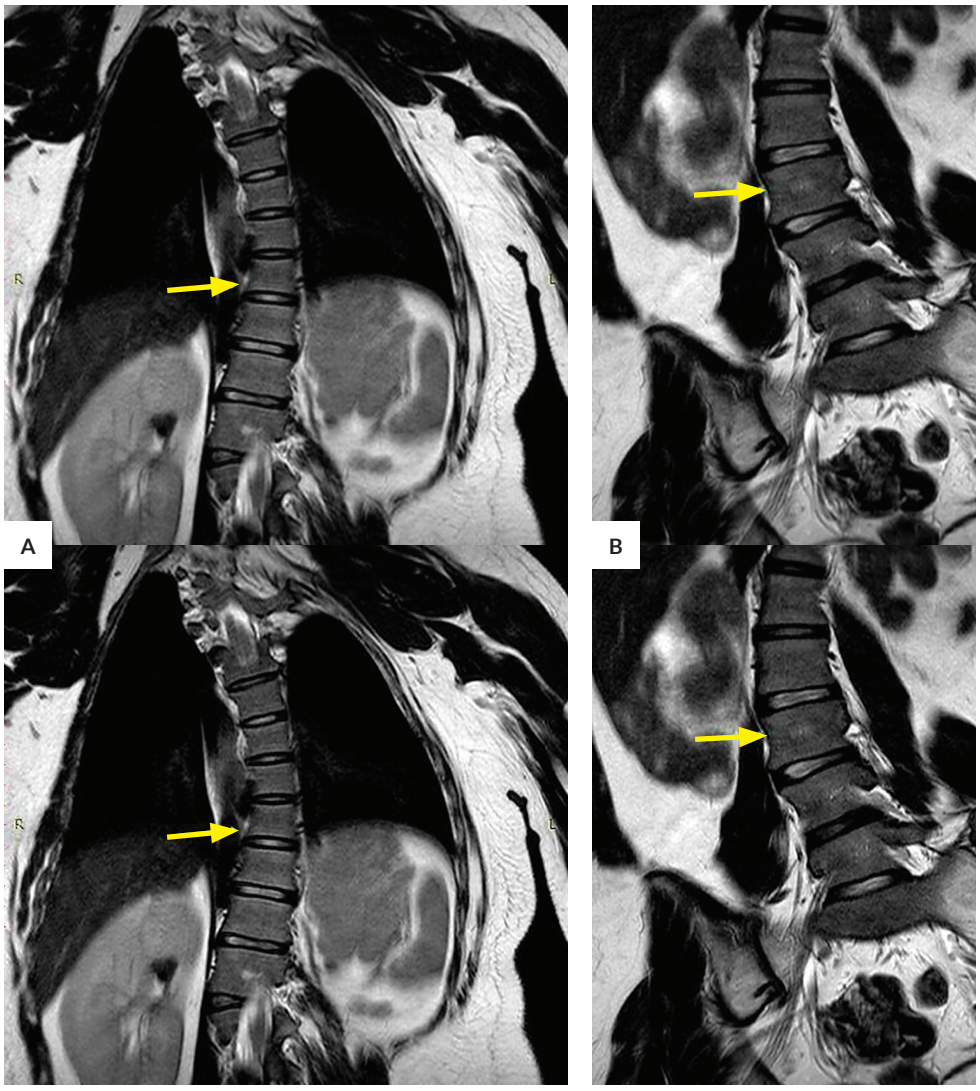

Figure 4. MRI of the spine showing **(A)** cervical and thoracolumbar dextroscoliosis and **(B)** thoracic levoscoliosis *(yellow arrows)*.

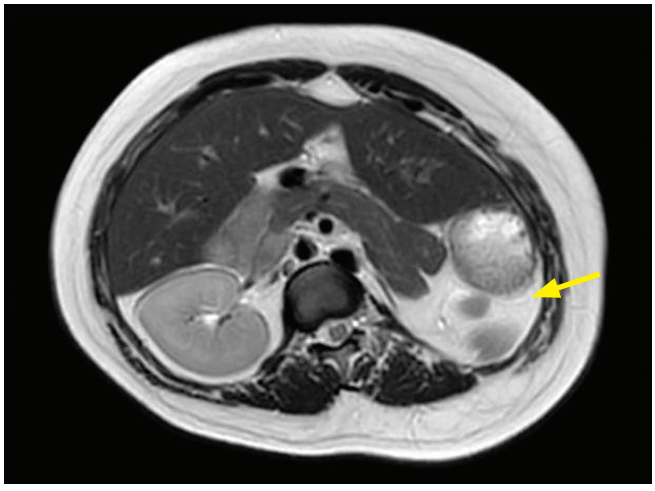

Figure 5. MRI of the abdomen showing an absent left kidney *(yellow arrow)*.

preloaded with a size 6.5 mm endotracheal tube (ETT) was then inserted through the oral cavity. There was no difficulty visualizing and approaching the glottic opening. After visualization of the carina, the ETT was advanced through the flexible fiberoptic scope and was secured at a depth of 18 cm.

Breath sounds were equal, and capnography showed a reading of 36 mmHg. Propofol 90 mg (1.6 mg/kg) and rocuronium 20 mg (0.37 mg/kg) were given intravenously for induction of anesthesia. Inhalational sevoflurane was started at 3% volume concentration with a gas mixture of 60% FIO_2. Pressure-controlled ventilation was used with the following settings: peak inspiratory pressure (PIP) of 20, PEEP of 5 and RR of 16. Remifentanil TCI was then decreased to 0.05 ng/ml. Sevoflurane was titrated to maintain MAC of 0.8 – 1.0. Dexamethasone 5 mg (0.09 mg/kg) and paracetamol 900 mg (16.67 mg/kg) were given per IV after induction. No surgical nor anesthetic complications were noted. Blood

loss was minimal. Intraoperative vital signs were stable with the following ranges: BP 110- 140/60 - 80, HR 80-100, SPO_2 100% $ETCO_2$ of 30-39 mmHg. Ondansetron 4 mg (0.07 mg/kg) per IV was given 30 minutes prior to the end of the surgery. The incision sites were infiltrated with 0.25% isobaric bupivacaine prior to suturing. At the end of the operation, sugammadex 200 mg (3.70 mg/kg) per IV was administered. Emergence was smooth with minimal hemodynamic variability. Patient was extubated fully awake and with intact airway reflexes. No bucking was observed during extubation.

Postoperatively, the patient was fully awake and comfortable. Vital signs were stable with BP of 130/70, HR of 90, RR of 18, and O_2 saturation of 100% at room air. Neurological examination was status quo with no new noted deficits. After an unremarkable postoperative course, she was discharged after two days.

As of this writing, she is on continuous medical management for infertility under the gynecological service, with no complications attributable to the surgery and anesthesia.

DISCUSSION

Operative laparoscopy typically requires general endotracheal anesthesia because tracheal intubation reduces the risk of bronchoaspiration, enables better control of ventilation, and allows for rigorous analysis of carbon dioxide.[8] Moreover, it eliminates the discomfort due to pneumoperitoneum and changes in positioning.[9]

Given the anticipated difficult airway, a neuraxial technique may be considered. There were reports of laparoscopic surgeries done under a neuraxial technique.[9,10] Due to her multiple vertebral fusion defects, her cervical and thoracolumbar dextroscoliosis and thoracic levoscoliosis, regional anesthesia is technically more difficult.[11,12] There is an increased risk of inadvertent dural puncture and unpredictable spread of local anesthetic within the anatomically distorted epidural space.[12] Spinal anesthesia would have a more predictable spread; however due to her short stature, there is an elevated risk for high block which might require emergency intubation.[11,12]

While proven feasible, systematic review and meta-analyses done showed that 3.4% of cases ultimately required anesthetic conversion.[13] An emergent airway management in the setting of a difficult airway is technically challenging and risky; hence, a planned and controlled airway management was preferable for this case.

Securing the patient's airway is made complicated by the unstable cervical spine and limited neck motion due to her cervical synostosis.[14] Even a low energy impact or injury during direct laryngoscopy, tracheal intubation, and positioning can induce a neurologic deficit.[15] Furthermore, flexion and extension of her neck should be avoided to limit further compression of her already herniating neural structures secondary to her Chiari malformation.[7] Awake video-assisted fiberoptic intubation was used because it allows the airway to be secured in a controlled, non-urgent manner without cervical manipulation.[11,12] Since this could be technically difficult, the otorhinolaryngology service was asked to stand by for an emergency tracheostomy.[12]

Sudden increases in intracranial pressure intraoperatively especially during induction, positioning, and extubation – like coughing and straining against the tube, should be prevented to avoid spinal cord damage.[7] Increases in intracranial pressure may aggravate the cerebellar herniation and may also transmit the higher pressure into the central canal causing syrinx formation.[7]

Pneumoperitoneum in laparoscopic procedures increases intracranial pressure, especially in abdominal pressures more than 15 mmHg.[7] This is attributed to increased intrathoracic pressure, impaired venous drainage of lumbar venous plexus, and late chemical mechanism due to increased $PaCO_2$.[7] To prevent this, abdominal pressure intraoperatively was maintained at 12 mmHg.

To achieve neuroprotection intraoperatively, mean arterial pressure was maintained more than 65 mmHg, systolic blood pressure was monitored and maintained at 20% of her baseline, and end tidal concentration was maintained at normocapnic levels. A balanced anesthetic technique using general anesthesia with sevoflurane and remifentanil was utilized. The latter's potent analgesic effect limited the use of sevoflurane– which can increase intracranial pressure due to its cerebral vasodilator effects at a minimum alveolar concentration (MAC) greater than 1.5.[16,17] Intraoperative MAC was maintained at 0.8 to 1.0. Moreover, remifentanil infusion attenuated the cardiovascular response during intubation and allowed for tracheal extubation without significantly delaying recovery from anesthesia.[18] This was evident during the patient's smooth extubation and emergence.

A possible restrictive respiratory defect due to the patient's scoliosis leads to reduced lung volumes, limited diaphragmatic excursion, and chest wall muscle inefficiency.[9] The use of pressure-controlled ventilation improves oxygenation and gas exchange in patients with decreased lung compliance – such as in scoliosis, while simultaneously maintaining peak airway pressures and reducing PIP.[19]

Lung protective ventilation using low tidal volume of 6-8 ml/kg with use of positive end expiratory pressures (PEEP) were also utilized. The use of PEEP increases functional residual capacity and improves the compliance of the lung by causing further distention of alveoli and small airways, recruiting collapsed alveoli, and preventing collapse of small airways and alveoli at end-expiration.[19] Due to the effects of pneumoperitoneum and Trendelenburg position on the lung compliance, the tidal volume was monitored closely, and adjustments were done accordingly to maintain the desired tidal volume and avoid hypoventilation.

In the presence of renal anomalies, such as the renal agenesis evident in this patient, it is prudent to avoid or adjust medications eliminated mainly by the kidney.[19] For renal protection, nephrotoxic agents such as non-steroidal anti-inflammatory drugs (NSAIDs) were avoided. Sevoflurane, which can cause nephrotoxicity due to its metabolite, was minimized by the addition of remifentanil.[20] Judicious fluid loading was done by using an isotonic fluid solution at 1-3 ml/kg/hr. Volume status and urine output were monitored closely to ensure adequate renal perfusion.

CONCLUSION

Klippel-Feil Syndrome poses an anesthetic challenge due to the inherent difficult airway indicated by its clinical triad. Preoperative evaluation and screening, not only of the airway and of the vertebral anomalies, but also for other associated systemic syndromes is vital as this allows anesthesiologists to carefully plan for successful management of these patients. Knowledge of the possible effects of these anomalies enables the anesthesiologist to prevent or mitigate their effects and allows the patient to withstand the effects of anesthesia and surgery.

Informed Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Statement of Authorship

Both authors certified fulfillment of ICMJE authorship criteria.

Author Disclosure

Both authors declared no conflicts of interest.

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