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CASE REPORT

ANAESTHETIC CHALLENGES IN A PEDIATRIC PATIENT WITH KLIPPEL FEIL SYNDROME UNDERGOING SURGERY IN PRONE POSITION

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ABSTRACT: Seven years old male child with Klippel Feil syndrome and Sprengel shoulder was posted for Woodwards procedure in prone position. Patients with this syndrome are vulnerable to cervical spinal cord injury and are at high risk for neurological injury not only during laryngoscopy and intubation but thereafter. Paediatric patients with cervical spine instability and limited range of motion are challenge to anaesthesiologists.

KEYWORDS: Klippel Feil syndrome, Pediatric, Prone position, Difficult airway.

KEYMESSAGES: Pediatric patients with Klippel-Feil syndrome may require general anaesthesia for correction of associated congenital anomalies or for surgical stabilization of the cervical spine. Manipulation of the neck during intubation and thereafter must be carefully controlled if neurological damages to be avoided.

INTRODUCTION: KFS is usually 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.[1] These patients pose anesthetic challenge when they come for surgery. The parent provided written informed consent for the authors to publish this report.

CASE HISTORY: Seven year old male child with Klippel Feil syndrome (KFS) and Sprengel shoulder was posted for Woodwards procedure in prone position. On examination, mouth opening was 4cms, MP IV, short neck with restricted neck mobility 10^0flexion and extension. Systemic examination revealed wide split 2nd heart sound. Lab investigations were normal. Echo heart: Congenital acyanotic heart disease OS ASD with Left to Right shunt. No PHT.

CT cervical spine: Segmentation and fusion anomalies: Bifida occulta at C2; C1 C2 C3 fusion anomaly; C5 –C6 fixed. Difficult airway trolley was kept ready, intravenous premedication with injection Atropine 0.03mg, Midazolam 0.5mg, Fentanyl 20µg, pre-oxygenated for 5minutes, induced with Propofol 35mg and mask ventilation was adequate so patient given injection Atracurium 10mg given, mask ventilated for 3mts with Oxygen, Nitrous oxide and Sevoflurane, on laryngoscopy it was Cormack III visualization of the epiglottis and vocal cords, intubated with 5mm un-cuffed endotracheal tube with stylet with head in neutral position.

Meticulous attention was given to positioning of the patient. To maintain head in neutral position, a suitable size ring and additional horse shoe shaped head rest was placed under the head to ensure immobilization of neck and to prevent endotracheal tube kinking. (Figure-1) Transverse bolsters were placed under the chest and pelvic girdle to ensure free movement of the diaphragm.

Surgery proceeded uneventfully. As a part of multimodal analgesia, Paravertebral block was given under vision by the surgeon with 0.25% Bupivacaine 1.5ml in each paravertebral space from C7

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to T5 and Diclofenac suppository 50mg, 12th hourly and residual neuromuscular blockade was reversed with atropine 0.02 mgkg-1 and neostigmine 0.05 mgkg.-1 The tracheal extubation done after resumption of spontaneous breathing and the return of good muscle tone in neutral position. Post-operatively the patient was monitored in the recovery room after which he was sent to the postoperative ward.

DISCUSSION: KFS becomes a challenge to anesthesiologists when patients present for surgeries which require prone position and maintenance of head in neutral position. Severe restriction of cervical motion from fusion and cervical instability leads to difficult intubation in pediatric population where awake fiber-optic intubation (FOI) is not an option,[1] the most critical moments include airway manipulation during laryngoscopy, tracheal intubation and positioning of the patient for the surgery.[2] The successful use of tracheal intubation with fiber-optic bronchoscopy[3] and of the laryngeal mask[4] has been reported. Fiber-optic intubation technique is considered the reliable technique.

Awake intubation using fiber-optic bronchoscope has been used successfully by other anesthetist,[5] but this technique is not possible in children and patient with behavioral problems, as the technique mandates patient cooperation. Laryngeal mask has been successfully used in a patient with KFS who developed difficult to ventilate after anesthetic induction.[5] Syncope attacks may be precipitated by sudden rotatory movements of the neck in patients with Klippel Feil syndrome. O'Conner and Moysa suggested that airway control can be temporarily lost after induction and LMA may be required to attain control of the airway.[6]

There was also a failure in fiber-optic intubation on two separate occasions.[7] We decided to do a conventional laryngoscopy without cervical extension and documentation of Cormack-Lehane score, as this patient is likely to come for further surgeries, due to the visualization Cormack-Lehane III of the vocal cords the patient was intubated.

Although difficult airways are prominent on scientific information on KFS with Sprengel's deformity we did not find references of pediatric patient this syndrome with atrial septal defect coming for Woodwards procedure in prone position. Prone positioning of patients during anesthesia is required to provide operative access for a wide variety of procedures. It is known to be associated with a number of physiological changes and complications, maintaining prone position requires an understanding of both issues.[8]

Excessive neck extension during positioning combined with the muscle relaxation of general anesthesia was blamed, even though it has occurred during tracheal intubation. Cervical spine dislocation injuries seems to be rare; two patients are described with pre-existing cervical spine dislocations who were nursed on a Stryker Frame and whose dislocations recurred when turned from supine to prone.[9] However, de novo dislocation has not been described.

Usually surgical treatment of Sprengel deformity and the omovertebral bone is recommended when the patient is between 3 and 7 years of age, because risk of brachial plexus injury from stretching or compression by the clavicle increases with age. (Figure-2) Early diagnosis and surgical treatment may prevent secondary neurological damage.[10]

Knowledge of the anatomical changes and adequate technical resources are paramount, especially in the case of orthopedic surgeries in prone position and surgical incision extended from C6 to T8. Manipulation of the neck or an attempt to extend the neck during laryngoscopy and

thereafter must be carefully controlled if neurological damage is to be avoided.[11] Contrary to what was expected, in our patient we did not find any difficulties to mask ventilation and tracheal intubation.

The non–availability of appropriate size flexo-metallic endotracheal tube increased the challenge of maintaining the airway patent in neutral position. An appropriate size horse shoe shaped support was given to forehead and maxilla, beneath the horse shoe shaped head rest another head ring was placed into which face was placed. Meticulous care was taken to maintain hemodynamic stability for Klippel-Feil syndrome patient with atrial septal defect in prone position.

Pediatric patients with Klippel-Feil syndrome may require general anaesthesia for correction of associated congenital anomalies or for surgical stabilization of the cervical spine. Manipulation of the neck during intubation and thereafter must be carefully controlled if neurological damages to be avoided.

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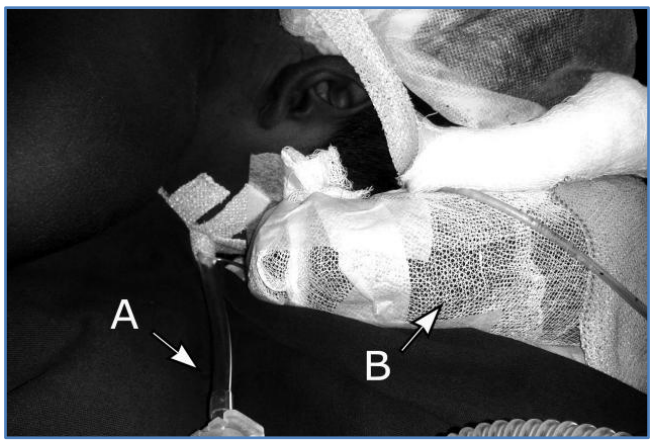

Fig. 1(a) Endotracheal tube. (b) Horse shoe shaped head rest

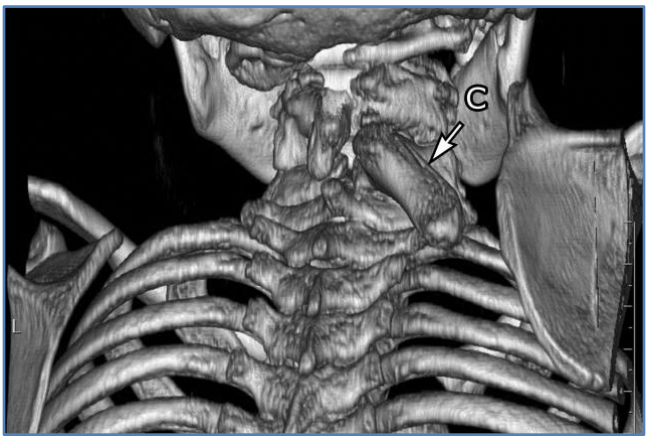

Fig. 2(c) Omovertebral body connecting scapula to vertebral lamina

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