Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2016; 4(1):22-24 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources) DOI: 10.36347/sjmcr.2016.v04i01.007

A Case of KlippelFeil Syndrome: Oral Intubation with the Help of Gum Elastic Bougie

Smitha KS[1], Manjunath Patil[1], Sreenivas Babu A[2]
[1]Registrar, [2]HOD,Department of Anesthesiology & Critical Care, The Bangalore Hospital, Bangalore, India

***Corresponding author**
Dr Smitha K S
Email: smi9685@gmail.com

Abstract: KlippelFeil syndrome is a rare disorder of skeletal system, presenting with fused cervical vertebrae, short neck, decreased or severe restriction in the neck movements and the associated anamolies like scoliosis, renal anomalies, cardiovascular abnormalities. They usually present to surgery and anesthesia for surgeries of scoliosis, renal anamolies and other general cases also. They come under anticipated difficult airway group. Neuraxial blockade in these patients is also unreliable due to abnormalities in the spine. For securing the airway, the technique of choice is fibre-optic bronchoscopy, however the other alternative methods are also successful many a times without much difficulty. Here, we report a case of KlippelFeil syndrome posted for Lapascopic ovarian cystectomy, intubated orally with the help of gum elastic Bougie.
Keywords: Cervical vertebrae,Intubation,KlippelFeil syndrome, Lapascopic, Scoliosis

INTRODUCTION

KlippelFeil syndrome(KFS)is a rare skeletal disorder, due to abnormal fusion of two or more cervical vertebrae. It is characterised by the triad of a short neck,alow posterior hair line and severe restriction of neck movements. The associated anamolies being polydactyl, scoliosis, renal anamolies, synkinesia and congenital heart disease(most commonly VSD).Incidence is 1 in 40,000 to 42,000 newborns worldwide [1].

The majority of these patients present to anesthesia for cervical spine correction, scoliosis surgery, renal surgeries, cleft lip/palate repair, Caesarian section or any other general surgeries [2, 3].

Due to the short neck, severe restriction of neck movements and cervical instability, these patients pose a challenge to Anesthesiologists in securing the airway. Regional anesthesia also poses a big challenge due to scoliosis and deformities of the spine. The effect of neuraxial blockade may be unreliable due to spinal deformity and compression of epidural or intrathecal space [4]. Awake fibre-optic bronchoscopy is considered as the gold standardtechnique for securing the airway [5]. The majority of the reports document the use of fibre-optic bronchoscopy to secure the airway. Also, there are reports of failure of fibre-optic bronchoscopy at two occasions [6].

At this moment, we report a case of Klippel-Feil syndrome posted for Laparoscopic ovarian Cystectomy, intubated with the aid of Bougie alone.

THE CASE

A 35 years old female patient came with history of lower abdominal pain since 5 days. She was diagnosed to be having right sided ovarian cyst and hence she was posted for laparoscopic ovarian cystectomy.

Pre-anesthetic examination revealed that the patient was non diabetic,not hypertensive with good exercise tolerance. She didn't have any history of asthma or any allergies. Her menstrual history was normal and had two normal vaginal deliveries of healthy kids. On examination,patient was short, moderately nourished and had short webbed neck with decreased range of flexion or extension of neck. Also she had low posterior hair line and 6 digits in her right hand. Airway examination revealed 3 finger mouth opening, Mallampati grade 3 with no flexion or extension of neck.Auscultation of chest and cardia were normal. Blood pressure 120/70mm of Hg and pulse rate 74/ minute.

Cervical spine X-ray lateral view revealedfusion of C2 C3 C4 and C5.Atlantoaxial joint was normal. Routine investigations done were in the normal range. Hemoglobin,blood sugars, RFT, chest x ray and ECG were normal.ECHO study was

normal.USG abdomen, done to look for any renal anamolies, was normal.

Patient was prescribed tablet Alprozolam 0.25 mg and tablet Ranitidine 150 mg orally night before the surgery and was kept nil by mouth for 8 hours.

On the day of surgery,after checking the anesthesia machine and suction apparatus,patient was shifted inside the operation theatre.18G cannula secured in the peripheral vein in the left hand and fluids started.Patient lied supine on the table comfortably.ECG,blood pressure and pulse oximeter monitors connected. Difficult airway cart was kept ready including the fibre optic bronchoscope. Injections Glycopyrolate0.2 mg, Midazolam 1mg and Fentanyl 100 micrograms given along with pre-oxygenation. Injection Propofolgiven in titrated doses to induce sleep.As mask ventilation was difficult due to inability to extend, oral Guedel's airway 3placed. After adequate depth of anesthesia with spontaneous ventilation being intact, a check laryngoscopy was done to visualise cords. But only epiglottis was visible. As we could mask ventilate with airway, we decided to give a trial of intubating with the help of Bougie after giving injection Succinylcholine. With Succinylcholine,we could see the posterior arytenoids. With the help of BURP manoeuvre, we passed the Bougie under the epiglottis in the direction of larynx, without the visualisation ofvocal cords. An endotracheal tube of 7 internal diameter rail-roaded through the Bougie, passed into the trachea and Bougie withdrawn. The cuff of the endotracheal tube inflated, auscultated for bilateral air entry and intra-tracheal placement confirmed with EtCO2 reading.Tracheal intubation was successful with the help of Bougie in the first attempt.

The patient was connected to anesthesia ventilator, with O2, N2O and Isofluraneinhalational anesthesia. Within ten minutes of Injection Scoline, the patient had respiratory attempts and hence Injection Vecuronium 6 mg given. Hemodynamics of the patient was stable. The surgery started which lasted for 30 minutes. 1litre of crystalloids transfused during the surgery. Neuromuscular blockade reversed with Injections Neostigmine and Glycopyrrolate. The patient obeyed commands and generated good tidal volume with regular breathing pattern. She was extubatedun eventfully and was observed in the recovery room for 3 hours. Vitals being stable throughout the recovery period, she was shifted to the ward.

DISCUSSION

Klippel-Feil syndrome was first described by Klippel and Feil in 1912 [7]. Along with fusedunstable cervical vertebrae, there might be fusionatAtlanto occipital joint leading to neurological damage. Syncopal attacks may occur by sudden rotatory movements of the neck. Our patient's C2 C3 C4 and C5 vertebrae were fused with stable Atlanto occipital and Atlanto axial joints.

The associated anamolies with the syndrome are scoliosis with fusion of ribs,occipito-cervical abnormalities leading to orthopaedic and neurological complications [8], polydactyl, deafness [9], renal anomalies [10], cardiovascular abnormalities[11, 12] (most commonly VSD) and craniosynostosis [13]. Our patient was polydactyl with 6 digits in her right hand, did not have any renal anomalies as USG abdomen was normal and ECHO was also normal.

KFS patients are considered under the anticipated difficult airway group, who pose a challenge for intubation, because of the short neck,inability to extend and the cervical spine instability. Awake Fibre-optic is considered as the technique of choice for the intubation, more conveniently, nasal route. However, there are reports of other options used.Oral intubation with anterior laryngeal pressure can be done on trial basis, however other modalities being on standby. Blind nasotracheal intubation[14]is one of the options, but this technique requires flexion and extension of neck which is contradicted in unstable cervical spine.

The intubating LMA can be used without much manipulation of the neck.But, there are concerns over the cervical pressures generated by the LMA which cause posterior displacement of the normal cervical spine. Hence, their use in unstable spine is controversial [15]. We gave a trial of oral intubation by giving injection Scoline, as we could ventilate the patient with mask. With the help of gum elastic Bougie, we could pass the endotracheal tube successfully.

According to a case series reported[16],a total of 10 patients underwent 11 procedures. Of the 10, 6 patients underwent tracheal intubations in which 5 were done in first or second attempt; only one patient required fibre-optic intubation. The remaining 4 had a LMA. The airways of KFS patients can be managed successfully in a variety of ways, often with little difficulty.

CONCLUSION

The airways of KFS patients are considered as anticipated difficult airway. Difficult airway cart has to be in place while anaesthetising these patients.Considering all the options available, fibre-optic intubation stands out as the gold standard. However, as mentioned earlier, other various options of intubations are successful at most times without much difficulty.

The difficult airway should be approached with caution to ensure favourable outcome. All it

includes is, a comprehensive preoperative examination and investigations, the availability of several alternative techniques, a difficult airway cart, a promptness to call for an expert help, the availability of the surgeons standing by to secure a surgical airway if required and a good amount of common sense with a calm mind.

REFERENCES

1. KlippelFeil syndromeavailable from http://ghr.nlm.nih.gov/condition/klippel-feil - syndrome.
2. Guille JT, Miller A, Bowen R, Forlin E, Caro PA; The natural history of the syndrome:clinical, roentgenographic and magnetic resonance imaging findings at adulthood. J PediatrOrthop., 1995; 15:617-626.
3. Wong J, El Beheiry H, Rampersaud YR, et al; Tranexamic acid reduces perioperative blood loss in adult patients having spinal fusion surgery. Anesth Analg., 2008;107:1479-86.
4. Dresner MR, Maclean AR; Anaesthesia for Caesarean section in a patient with Klippel-Feil syndrome. Anaesthesia, 1995; 50:807-09.
5. Daum RE, Jones DJ; Fibreoptic intubation in Klippel-Feil syndrome. Anaesthesia, 1988; 43: 18-21.
6. Asai T, Shingu K; Difficulty in advancing a tracheal tube over a fibreoptic bronchoscope. Br J Anaesth., 2004; 92(6):870-81.
7. Naguib M, Farag H, Ibrahim AEW; Anaesthetic considerationsin Klippel- Feil Syndrome. Can AnaesthSoc J., 1986; 33:66-70.
8. Hensinger RN, Lang JE, MacEwen GD; Klippel-FeilSyndrome; a constellation of associated of anomalies. J Bone Joint Surg Am., 1974;56(6): 1246-53.
9. Dubey SP, Ghosh LM; Klippel-Feil Syndrome with congenital conductive deafness: report and review of literature.Int JPediatrOtorhinolaryngol.,1993; 25(1-3): 201-08.
10. Ramsey J, Bliznak J; Klippel-Feil Syndrome with renalagenesisand other anomalies.AnRoentgenol Radium TherNucl Med.,1971; 113(3): 460-63.
11. Algom M, Schlesinger Z; Prolapse of the mitral valve in Klippel-Feil syndrome. Chest, 1981; 70: 127-28.
12. Falk RH, Mackinnon J; Klippel-Feil syndrome associated with aortic coarctation. Br Heart J., 1976; 38: 1220-21.
13. Chattopadhyay A, Shah AM, Kher A et al; Craniosynostosis and Klippel Feil syndrome: Its association. Indian J Paediatr.,1996; 63(6): 819-22.
14. Crosby ET, Lui A; The adult Cervical Spine: implications forairway management. Can J Anaesth., 1990; 37: 77-93.
15. Keller C, Brimacombe J, Keller K; Pressures exerted againstthe cervical vertebrae by the standard and intubating laryngealmask airways. AnesthAnalg., 1999; 89: 1296-1300.
16. Stallmer ML, Vanaharam V, Mashour GA; Congenital cervical spine fusion and airway management: a case series of Klippel-Feil syndrome. Journal of clinical anesthesia, 2008; 20(6):447-451.