

CASE REPORT

ANESTHETIC CHALLENGES FACED IN A CHILD WITH TREACHER-COLLINS SYNDROME

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ABSTRACT: Anesthesiologists come across pediatric patients with rare diseases and syndromes scheduled for various operative interventions. Treacher Collins syndrome is a rare genetic disorder characterized by craniofacial deformities, the incidence being 1 in 40, 000 - 70, 000 births¹⁻³. Treacher Collins syndrome (TCS) poses serious problem in securing and maintaining airway due to facial deformity. Difficulty in intubation increases as the patient's age increases. It requires meticulous planning and assessment of the airway prior to each anesthetic technique. Here we describe and discuss successful anesthetic management of an 8 year old boy posted for canaloplasty of the right ear.

KEYWORDS: Treacher Collin's syndrome, Difficult airway, Micrognathia.

INTRODUCTION: Treacher-Collins syndrome is a rare disorder of craniofacial development which results in mandibulofacial dysostosis. It is a congenital malformation of the first and second bronchial arches inherited as autosomal dominant trait with a variable expressivity.

Most of the patients with TCS present with the following classic facial features: down-sloping palpebral fissures, colobomata of the lower eyelid, scanty lower eyelashes, malar hypoplasia, and micrognathia or retrognathia.

The patients with TCS have a serious problem in mask ventilation, securing and maintaining airway in the perioperative period. Retrognathia, macroglossia and other associated abnormalities result in limited mouth opening, reduced extension of the head on the neck, hypoplastic mandible and limited forward movement of hyoid.

During the postoperative period, pharyngeal and laryngeal edema may result in respiratory distress and sudden death which have been reported in the literature.

We report a case of general anaesthesia in a child with TCS who was scheduled for canaloplasty of the right ear.

CASE REPORT: An 8-year-old boy weighing about 20kg, diagnosed as Treacher-Collins syndrome was scheduled for canaloplasty of the right ear. On evaluation the patient was found to have hypoplasia of facial bones, down sloping palpebral fissures, coloboma of the lower eyelids, scanty lower eye lashes and micrognathia and retrognathia. The child had narrowing of auditory canals on both sides, for which he was taken up for canaloplasty of right ear. Airway assessment revealed adequate mouth opening with Mallampatti class 4 airway. Neck movements and spine were normal. All investigations and systemic examination were normal.

In view of the difficult airway, tracheostomy consent was obtained. A difficult airway cart was kept ready including LMA, tracheostomy set and emergency cricothyroidotomy set. The child was fasted for six hours. As age was a limitation for awake intubation, we planned for smooth induction

CASE REPORT

with a deeper plane of anesthesia ensuring adequate ventilation and preventing trauma to the airway. Child was premedicated with glycopyrrolate 0.2mg IV to reduce the secretions. With standard Monitoring and preoxygenation anaesthesia was induced with incremental dose of sevoflurane & IV propofol 1mg.kg⁻¹. Initially mask ventilation seemed to be difficult due to a poor mask fit but improved to some extent after an oropharyngeal airway insertion and gauze packing of the space between the mask and the cheek. Adequate ventilation was achieved after applying jaw thrust. After induction, a gentle laryngoscopy with Macintosh blade showed a Class IV of glottis visualization as per Cormack Lehane classification.

With incremental doses of sevoflurane, appropriate depth of anesthesia was achieved. With the child's head turned slightly to the left and the assistant was asked to give a very good backward upward right-ward pressure (BURP). With this maneuver laryngoscopy revealed a Cormack Lehane of glottis visualization as Class III. Intubation was then accomplished with no.6 cuffed endotracheal tube with the help of a stylet. Correct placement of the tracheal tube was confirmed. Further anesthesia was maintained with N₂O+ O₂+ sevoflurane and atracurium with fentanyl 2µg.kg⁻¹ IV for analgesia. Adequate oxygenation and good hemodynamic stability was maintained through the surgery. Dexamethasone 2mg was also given intravenously. At the end of the surgery, the neuromuscular block was reversed with neostigmine 0.05mg.kg⁻¹ and glycopyrrolate 0.02mg.kg⁻¹. Extubation was carried out when the child got awake and thereafter shifted to recovery ward for further postoperative monitoring. The patient was discharged after 7 days without any complications.

DISCUSSION: The patients with TCS, posted for various surgeries, pose a problem to the anesthetists with regards to their airway management. It may be due to relative macroglossia as a consequence of skeletal abnormalities. The abnormalities which are associated with TCS may be limited mouth opening, reduced extension of the head on the neck, a hypoplastic mandible and a limited forward movement of the hyoid. Often, multiple mechanisms may be present in an individual case³⁻⁶.

Our patient presented with classical features of Treacher-Collins syndrome with significant airway distortion, because of which we had expected difficulty in maintaining airway as well as difficult tracheal intubation.

Awake fiberoptic intubation can be uncomfortable and stressful for the patient and it requires expertise and patient's cooperation. Retrograde tracheal intubation provides higher success rate. It is traumatic & painful especially in pediatric patients⁶⁻⁸. Bullard's Laryngoscope has also been tried for intubation⁹.

These difficulties can be easily overcome by using LMA & ILMA (Intubating LMA). However, the pediatric patients with Treacher-Collins syndrome have the posteriorly protruded tongue which displaces the LMA, makes the glottis move considerably anterior and interfere with the attempts to enter the trachea with a bougie. Further downward displacement of the epiglottis can also impair the intubation technique through LMA. Similar problems are encountered while attempting light wand guided intubation¹⁰⁻¹³.

Recently airway management of children with Treacher Collins syndrome (TCS) has been reviewed by Hosking, j.et al.in 240anesthetics. MCL grade increased with increasing age (**P** = 0.007)¹⁴ Considering the pros & cons of the various techniques, and keeping difficult airway trolley & tracheostomy kit as standby, we tried attempting conventional approach with direct laryngoscopy

CASE REPORT

with suitable modifications. We avoided the use of sedatives that depress respiration preoperatively. Intraoperatively we achieved smooth & deeper plane of anesthesia by inhalational agents. Patient was kept breathing spontaneously and deeper plane of anesthesia was achieved to provide adequate time to attempt intubation. The forward lift of both the angles of the mandible by an assistant facilitated ventilation to a great extent as retrognathia is main constraint in TCS patients. And finally intubation was facilitated by a straight blade, lateral approach, very good backward upward and rightward pressure (BURP) by an assistant which made Cormack Lehane of glottis visualization as Class III.

The straight blade causes minimal compression on the soft tissues of the large sized tongue thus allowing an equal distribution of tongue tissue on either side of the blade and making a clear central path for proper visualization of the glottis and intubation. Improved view by extension of the head is possible with use of straight blade. Lateral placement of the blade bypasses the tongue^{15,16}.

Anticipation of a difficult airway and intubation is very crucial in successful management of these patients. We employed modified conventional technique using lateral approach to laryngoscopy, forward lifting of both the angles of the mandible and BURP maneuver to intubate our patient.

REFERENCES:

1. Harrison MS. The Treacher Collins-Franceschetti syndrome. *J Laryngol Otol.* 1957; 71:597-604.
2. Victor C Baum, Jennifer E O'Flaherty, Anesthesia for genetic metabolic dysmorphic syndromes of childhood. *Lippincott Williams.*2006; 369-70
3. Tessier P. Anatomical classification of facial, cranio-facial and latero-facial clefts. *J Maxillofac Surg*1976; 4:69-75.
4. Samsoun GL, Young JR. Difficult tracheal intubation: a retrospective study. *Anesthesia.* 1987 May; 42(5): 487-490.
5. Splendor A, Jabs EW, Passos-Bueno MR. Screening of TCOF1 in patients from different populations: Confirmation of mutational hot spots and identification of a novel missense mutation that suggests an important functional domain in the protein treacle. *J Med Genet* 2002; 39:493-5.
6. Tsaun GR, Liaw WJ, Wong CS, Yu FK, Hwang JJ, Chen SG, Ho ST. Treacher-Collin's syndrome and translaryngeal guided intubation--case report. *Acta Anaesthesiol Sin* 1994 Sept;32(3):223-8
7. Bellhouse CP, Dore C. Criteria for estimating likelihood of difficulty of endotracheal intubation with Macintosh laryngoscope. *Anesth Intensive Care.* 1988; 16: 329-37.
8. Mallampati SR, Gatt SP, Gugino LD, et al. A clinical sign to predict a difficult tracheal intubation-a prospective study. *Canadian Journal of Anesthesia.* 1985; 32:429-34.
9. Agrawal S, Asthana V, Sharma JP, Sharma UC, Meher R. Alternative intubation technique in a case of Treacher-Collins syndrome. *The Internet J Anesthesiology.* 2006; 11:1-8.
10. Harea J. The Bullard laryngoscope proves to be useful in difficult intubations in children with the Treacher Collins syndrome. *Anesth Analg.* 2004; 98:1815-6; author reply 1816
11. Takita K, Kobayashi S, Kozu M, Morimoto Y, Kemmotsu O. Successes and failures with the laryngeal mask airway (LMA) in patients with the Treacher Collins syndrome - a case series. *Can J Anaesth.* 2003; 50:969-70.
12. Ebata T, Nishiki S, Masuda A, Amaha K. Anaesthesia for the Treacher Collins syndrome by using a laryngeal mask airway. *Can J Anaesth.* 1991 Nov; 38(8):1043-45

CASE REPORT

13. S. Hooda S, Nandini S, Sekhri C. Difficult airway management in a maxillofacial and cervical abnormality with an intubating laryngeal mask airway. *J Oral Maxillofac Surg.* 2004 Apr; 62(4): 510-13.
14. Hosking J, Zoanetti D, Carlyle A, Anderson P, Costi D. *Paediatr Anaesth.* 2012 Aug; 22(8):752-8. doi: 10.1111/j.1460-9592.2012.03829.x. Epub 2012 Mar 7.
15. Handler SD, Keon TP. Difficult laryngoscopy/intubation the child with mandibular hypoplasia. *Annals of Otolaryngology, Rhinology, Laryngology.* 1983; 92:401-4.
16. Diaz ZH, Guarisco JL, LeJeune FE Jr. A modified tubular pharyngo-laryngoscope for difficult pediatric laryngoscopy. *Anesthesiology.*1990; 73:357-8.



Fig. 1: Frontal view of the face showing micrognathia and other typical facies of Treacher Collins syndrome (TCS)



Lateral view(TCS)

CASE REPORT



Fig. 3: Intubation with no. 6

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