

Case Report

Anaesthetic management of a case of Treacher - Collins syndrome

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Abstract

Treacher-Collins syndrome (TCS) is a rare congenital disease known to be associated with a difficult airway and represents some of the most hazardous and difficult challenges that anaesthetists may encounter during their entire practice of anesthesia. Successful anaesthetic management of a case with Treacher-Collins syndrome posted for laparoscopic cholecystectomy under general anaesthesia is presented in this report.

Key Notes: Treacher-collins syndrome, anaesthetic management

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Introduction

Treacher Collins syndrome is referred to as mandibulofacial dysostosis, characterized by maxillary, zygomatic, and mandibular hypoplasia and known to be associated with difficult intubation. It is a congenital malformation of first and second bronchial arch, inherited as autosomal dominant trait.¹The basic etiology is obscure. IdaMaan in 1943 mentioned that a disturbance in division and development of the mesodermal bone tissue at fifthweek of foetal life probably initiates this syndrome.² The syndrome consists of congenital and familial deformities of the ear, eyes, maxilla and mandible. It is often associated with deafness due to meatal atresia and malformation of the middle and inner ear. Coloboma of the lower eyelids, scanty lower eye lashes, microtia with hearing loss and micrognathia and retrognathia^{3,4} may be present. During the post operative period, pharyngeal and laryngeal edema may develop. Even respiratory distress and sudden death has been reported⁴

Case Report

A female patient age of 23 years, weighing about 65 kg was scheduled for laparoscopic cholecystectomy in a private hospital. On pre-anaesthetic evaluation the patient was found to have hypoplasia of facial bones (mandible, maxilla and cheek), micrognathia and nasal obstruction gross deviated nasal septum. These features raised the suspicion that it is a rare case of Treacher Collins syndrome .On airway assessment mouth opening was found to be less then 3 cm and Mallampati Grading Class-4. (Fig 1, 2).



Fig 1

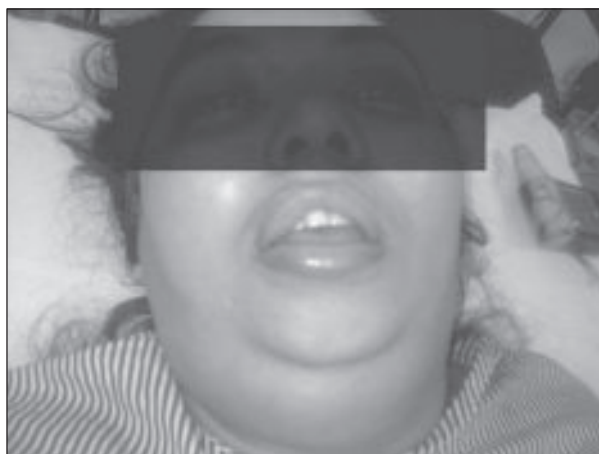


Fig 2

Neck movements and spine were normal. Preoperative blood investigations showed Hb- 11.5 gm%, No abnormalities were detected in other investigations.

Relatives were informed about the possibility of the difficult airway and on the event of failed intubation tracheostomy consent was taken. A trolley for difficult airway was kept ready including LMA, and tracheostomy set.

The patient was kept nil by mouth for six hours. We planned to go for smooth induction with a deeper plane of anesthesia, avoiding hypoventilation and trauma to the airway. The patient was premedicated with atropine 0.6mg IV to reduce the secretions. Sedatives were avoided as we anticipated a difficult airway. Dexamethasone 0.2mg.kg⁻¹ IV was given. The patient was preoxygenated with 100% oxygen for five minutes. Induction was done with IV propofol 2 mg.kg⁻¹ with fentanyl 2 µg/kg⁻¹ and suxamethonium 2 mg kg⁻¹. Initially mask ventilation seemed to be difficult due to 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. But even with these we could not ventilate adequately. Then one assistant was asked to lift forward both the angles of the jaw, and only then the patient could be ventilated. Now taking the patient in deeper plane, one assistant was asked to give a very good backward upward rightward pressure (BURP) on the airway. With this maneuver, Cormack Lehane classification of glottis visualization was Class III⁵ during laryngoscopy. Now we were able to intubate with a 7mm cuffed endotracheal tube with the help of a stylet. Later the tube was secured properly and the patient was handed over to the surgeons. Further anaesthesia was maintained with N₂O + O₂ + halothane and rocuronium 0.6mgkg⁻¹ with a supplementation of fentanyl 2µg.kg⁻¹ IV for analgesia. The patient was monitored with pulse oximetry, ETCO₂, NIBP, and precordial stethoscope for heart rate throughout the surgical period which lasted for about 45 minutes and rest of the procedure was uneventful. At the end of the surgery, the patient was reversed with neostigmine 0.05mg.kg⁻¹ and atropine 0.02mg.kg⁻¹. A smooth extubation was done. The patient was kept under observation and the

postoperative period was uneventful. The patient was discharged from hospital after 2 days without any complication.

Discussion

Patients with Treacher-Collins syndrome present a serious problem to anaesthetists in maintaining their airway, as upper airway obstruction and difficult tracheal intubation due to severe facial deformity. Because of retrognathia, the airway management of these patients is often challenging.

Another cause for difficult intubation in such cases is due to relative macroglossia as a consequence of skeletal abnormalities. This reduces the space available for manipulation and insertion of the endotracheal tube (ETT). The associated abnormalities like limited mouth opening, reduced extension of the head on the neck, hypoplastic mandible, limited forward movement of hyoid may be present.

Treacher-Collins syndrome is caused by a defective protein called treache. More than half of the cases are thought to be due to new mutations. Because there is no family history of the disease, the condition may greatly vary in severity from generation to generation.²

Our patient a case of Treacher- Collins syndrome with most of the features of significant airway distortion. That's why we had expected difficulty in maintaining airway as well as difficult tracheal intubation. Various techniques have been described in management of such patients. These include; direct laryngoscopy, intubation with a flexible fiberoptic bronchoscope, lightwand, laryngeal mask airway, retrograde intubation technique and tracheotomy can also be employed.⁵

In our case we make three important modifications of the technique. These were:

1. We used a short acting muscle relaxation which give adequate relaxation but give less time to attempt intubation.^{6,7} If we fail it is easy to recovered.
2. The forward lift of both the angles of the mandible by an assistant to overcome the main cause of difficult ventilation in TCS, the retrognathia.
3. And finally intubation was facilitated by a very good backward upward and rightward pressure (BURP) by an assistant.

Anesthesia is a field of challenges, especially when you encounter difficult to ventilate and difficult intubation scenario.⁵ Hence every anaesthetist should be well prepared with the various techniques of the difficult airway algorithm. This case of Treacher-Collins syndrome illustrates how a modified conventional approach can still be a very good and gold standard approach when other newer techniques are not available.

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