Crafting a Solution: An Impromptu Approach to Intubation in a Patient With Treacher-Collins Syndrome, Utilizing Video-Laryngoscopy and a Custom J-Shape Stylet

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Abstract- Treacher-Collins syndrome (TCS) is a rare congenital disease known to be associated with a difficult airway and is challenging for anesthesiologists practicing pediatric anesthesia. Many features of the syndrome are predictors of a difficult airway, including limited mouth opening, mandibular hypoplasia, relative macroglossia, glossoptosis, and obstructive sleep apnea. In this report, we share our experience with a TCS patient who presented for adenotonsillectomy and underwent successful intubation using video laryngoscopy with a revised J-shaped stylet.

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Introduction

Treacher-Collins Syndrome (TCS), also known as mandibulofacial dysostosis or zygotemporoauromandibular dysplasia, is an autosomal-dominant disorder with variable penetrance and no gender predilection, characterized by a constellation of several craniofacial malformations. Its incidence is estimated at 1 in 50,000 live births. Approximately 60% of cases are due to spontaneous or de novo mutations, while 40% are family-specific mutations. It is named after the British ophthalmologist Edward Treacher Collins (1).

The disorder displays an intricate underlying dysmorphology. Affected patients may suffer life-threatening airway complications and functional difficulties involving sight, hearing, speech, and feeding. It is a disorder of neural crest cell proliferation involving the development of the first and second branchial arches. Abnormalities are bilateral, symmetrical, and restricted to the head and neck (1,2).

TCS consists of deformities, in various degrees, of the eyes, ears, maxilla, and mandible. There is a wide range of ophthalmic anomalies, including Periorbital anomalies, coloboma, lacrimal system dysfunction, and refractive/vision problems. As for ears, it can be associated with deafness due to meatal atresia and malformation of the outer, middle, and inner ear (3).

Midface anomalies consist of the Pierre Robin sequence (PRS) triad (micrognathia, glossoptosis, airway obstruction-because of pronounced retrognathia). The mandible, however, is not only hypoplastic but also malformed (4).

A cleft palate is present in 40% of cases. Dental anomalies, Malocclusion, and Choanal atresia are some of the other anomalies that could also be present. Renal anomalies and congenital heart disease may be coexisting; however, these are not hallmark features and are not always consistent findings in affected patients. TCS patients usually have normal cognition and

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intelligence (4,5).

Obstructive sleep apnea (OSA) is often present and results from multilevel airway obstruction (1). Limited mouth opening, reduced extension of the head and neck, hypoplastic mandible, and limited forward movement of the hyoid are features that make airway management very challenging in TCS. Often, multiple abnormalities of varying degrees may be present in an individual case, making the management of each case of TCS unique (6,7). Also, during the postoperative period, pharyngeal and laryngeal edema may develop. Even respiratory distress and sudden death have been reported (8).

Case Report

A 6-year-old male child of Treacher-Collins syndrome weighing 20 kg was posted for adenotonsillectomy in the Anesthesiology department at Imam Khomeini Hospital Complex (IKHC). He had a history of Laryngomalacia at birth, bilateral hearing problems, recurrent URTI, and severe OSA with snoring. He had no previous surgical history.

Initial assessment revealed pectus excavatus, hypoplasia of facial bones (mandible, maxilla, and zygomatic), down-sloping palpebral fissures, scanty lower eyelashes, micrognathia, and retrognathia (Figure 1). On airway assessment, the mouth opening was adequate with a class 4 Mallampati airway. Neck movements were full range of motion without restrictions. The preoperative cardiac evaluation revealed that the patient had PFO and a mild MVP. Pre-OP lab data were within normal ranges. The chest X-ray and ECG were also normal. In the context of a difficult airway, a parent meeting was held, during which they were educated on the airway management risks associated with their child, including the possibility of emergency tracheostomy. A written informed consent was obtained.

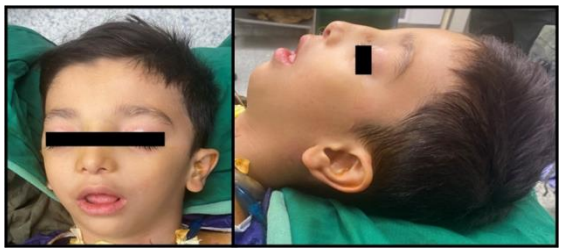

Figure 1. Front view: Treacher-Collins syndrome patient showing hypoplasia of facial bones, down-sloping palpebral fissures, and scanty lower eyelashes. Lateral view, showing retrognathia

On the operation day, sedatives were avoided when transferring the child to the operating theatre as we were anticipating a difficult airway. The initial anesthesia plan was Nasal fiberoptic bronchoscopy (FOB) and intubation with emphasis on preserving spontaneous ventilation. However, other difficult airway modalities were also present in the room. The surgical team was also briefed and ready in case of an emergency surgical airway rise. The patient's initial vital signs were: BP: 90/60 mmHg, heart rate 100 beats/min, and SpO2 95% in room air.

After standard monitoring, the patient was positioned into a proper sniffing position and preoxygenated with tidal volume breaths using 100% oxygen for 3 minutes. The patient received 20 mcg of Fentanyl as premedication. Nasal preparation was done using 10% topical viscous lidocaine gel combined with phenylephrine shrink. Induction began with 8% sevoflurane and continued with decremental doses to maintain spontaneous ventilation. The primary concerns were to maintain spontaneous ventilation while avoiding desaturation and airway trauma. The mask seal was acceptable, but due to severe OSA and relative macroglossia, oropharyngeal airway (OPA) combined with the jaw thrust maneuver was required for adequate ventilation. Despite spontaneous breathing, ventilation remained challenging throughout the airway management as sevoflurane aggravated the OSA and worsened airway patency. This became more problematic later on with a fiber optic intubation attempt.

After the appropriate depth of anesthesia was reached, FOB started with a 4.0x65 Flexible Intubation Videoendoscope (FIVE 4.0) from KARL STORZ. As the scope approached the nasopharynx, minor mucosal bleeding and secretions began to obscure the view, while the large tongue base obstructed the pathway and limited maneuverability during the procedure. This slowed the intubation attempt, rendering it infeasible within a reasonable time frame. It was decided to abandon FOB before the critical desaturation, and mask ventilation with sevoflurane was reinitiated.

As the next step in the plan, a UEscope VL300M video-laryngoscope with a Macintosh #2 blade and the included standard rigid stylet was used for oral endotracheal intubation. A #5 cuffed endotracheal tube from GreenMed was selected primarily based on age. Gentle video laryngoscopy was performed, revealing a Class III Cormack-Lehane classification; however, attempts to pass the tube were unsuccessful. The Backward-Upward-Rightward Pressure (BURP) maneuver did not improve the Cormack-Lehane

classification nor make the tube passage feasible. The standard rigid stylet used did not provide enough dexterity to reach the glottis, considering the anterior placement of the glottis and the large body of the tongue. Due to the start of the desaturation attempt, the patient was mask-ventilated. Adequacy of ventilation and feasibility were discussed with the team at this point. Even though ventilation required an Oropharyngeal airway and jaw thrust, it stayed consistently adequate. Given the expertise of a senior team member, other modalities, like intubating supraglottic airway devices (SGAs), were also discussed. However, the decision was made to proceed with video laryngoscopy using a revised stylet. During the third attempt, video laryngoscopy was employed with a manually shaped J-shaped stylet (Figure 2). This time, the stylet had better dexterity in reaching the anterior glottis, as it was shaped to better fit the patient's anatomy. Meanwhile, the lateral deviation of the proximal part helped maintain the view at all times. Eventually, the patient was successfully intubated using a No. 5 spiral cuffed endotracheal tube with the aforementioned stylet. Tube placement was confirmed by auscultation and capnography, then secured by tape. Vital signs remained within normal range, and there was no trauma to the airway or vocal cords.

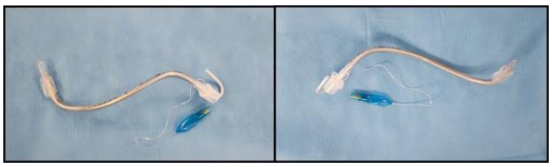

Figure 2. No. 5 cuffed spiral endotracheal tube equipped with manually J-shaped stylet

Anesthesia was maintained with sevoflurane, without the use of any muscle relaxant. 4 mg of dexamethasone was administered to alleviate any airway inflammation that may have occurred. The patient was monitored with pulse oximetry, ETCO2, NIBP, ECG, and Bispectral index throughout the surgical period, which lasted for about two hours. Considering the severe OSA patient received elective tracheostomy at the end of the tonsillectomy. The patient was closely monitored during the uneventful postoperative period and was transferred to the ICU awake, with spontaneous breathing.

Discussion

TCS patients are commonly scheduled for craniofacial surgery (33%), ENT surgery (21%), computed tomography (CT) scans (18%), dental procedures (9%), and, less commonly, vascular access, general surgery, and neurosurgical procedures (2%).

The main consideration for the anesthesiologist is the expectation of and preparation for difficult airway management. There is wide variability in the phenotypical presentation and the severity of facial dysmorphisms in TCS patients. Many features of Treacher-Collins Syndrome are predictors of difficult airway, including limited mouth opening, mandibular hypoplasia, relative macroglossia, glossoptosis, and OSA (1,9). Direct laryngoscopy becomes more difficult with increasing age secondary to dysmorphic facial growth. This contrasts with children in the Pierre Robin sequence, who get easier to intubate as they grow older (8).

It is beneficial to pay extra attention to the history of problems at birth, signs, and the degree of airway obstruction, as well as whether it is aggravated in different positions. A careful study of CT scans and polysomnography can provide valuable information. Another interesting modality is flexible nasolaryngoscopy at the bedside to identify the patency of choanae, degree of oropharyngeal obstruction, tongue base collapse, and laryngeal abnormalities.

Firstly, mask ventilation in TCS can be challenging. Anesthesiologists should anticipate a poor mask seal due to the dismorphology of mid-face elements, and ventilation may be difficult due to multilevel upper airway obstruction. Modalities like nasopharyngeal airways (NPA), oropharyngeal airways (OPA), and supraglottic airways (SGAs) combined with Airway maneuvers (Jaw thrust) may be required. It is advisable to have two experienced providers for bag-mask ventilation. Maintaining spontaneous breathing is of utmost importance.

Direct laryngoscopy will be difficult in these patients, and other methods for intubation should be considered. Fiberoptic bronchoscopy, Video laryngoscopy, Intubating LMAs, Lightwands, and retrograde intubation have all been tried and reported successful in TCS (10). Last but not least, emergency front-of-neck airways (Tracheostomy, cricothyroidotomy) should be available and discussed pre-OP with both the surgical team and parents.

Due to variable penetrance and a wide degree of deformities, every TCS patient should be considered a unique challenge. Meticulous adherence to airway management principles, preparation of different difficult airway modalities, and a probable Cannot Intubate, Cannot Oxygenate (CICO) scenario should be considered for every case.

We believe that what made our case successful was being prepared for it, having a clearly defined and communicated anesthetic plan. When there are multiple choices within a plan, the anesthesiologist's expertise, familiarity with equipment, and clinical vision, combined with the accompanying team experience, should be the determining factors in formulating the plan. The newer or more advanced modalities are not necessarily the best ones. Trusting the intuition of our senior anesthesiologist for choices within the plan showed that less common methods or even a modified conventional method can still achieve the desired outcome when an anesthetic plan encounters difficulties or advanced airway management modalities are simply unavailable. This emphasizes the importance of an anesthesiologist being experienced and comfortable with various methods of airway management. We hope that our shared experience becomes helpful in managing similar cases.

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