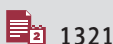


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Anesthetic Management of Treacher Collins Syndrome in an Outpatient Surgical Center

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Columbus, OH, USA**Corresponding Author:** Jarrett A. Heard, e-mail: Jarrett.Heard@osumc.edu
Conflict of interest: None declared**Patient:** Male, 15-year-old
Final Diagnosis: Treacher Collins syndrome
Symptoms: Difficult airway management
Medication: —
Clinical Procedure: —
Specialty: Anesthesiology**Objective:** Rare disease**Background:** Treacher Collins syndrome is a rare autosomal dominant disorder characterized by micrognathia and abnormal development of the zygomatic arch, which may result in significant upper airway obstruction. As patients who have it age, their upper airway obstruction may worsen. Therefore, they typically require several surgeries throughout their lives to correct specific facial abnormalities. Anesthetic and airway management of patients with Treacher Collins syndrome can be challenging for anesthesia providers, especially in ambulatory settings.**Case Report:** A 15-year-old patient with Treacher Collins syndrome presented to our outpatient surgery center for midface fat grafting. He had undergone multiple surgical procedures at Nationwide Children's Hospital, which is affiliated with The Ohio State University Wexner Medical Center. A decision was made to proceed with the grafting surgery after: (1) the literature was thoroughly reviewed; (2) multidisciplinary planning had been done utilizing our comprehensive preoperative screening and assessment process; (3) the scope of care at our ambulatory surgery center, the patient's medical history, and relevant airway notes had been reviewed; (4) the case was discussed with the surgeon; and (5) relevant images of the patient had been gathered. Evaluation of the patient's airway on the day of surgery was reassuring and a plan for managing a potentially difficult airway had been developed. After anesthetic induction, mask ventilation without adjuvants was successful. Video and direct laryngoscopy (for purposes of education) revealed grade 1 views. Supraglottic airway device placement resulted in an effective seal and the remainder of the surgery and the patient's subsequent course were uneventful.**Conclusions:** Improved airway approaches, combined with thorough preoperative screening and multidisciplinary planning and communication, may make it possible to perform ambulatory surgery on patients with Treacher Collins syndrome, whose condition typically represents a significant challenge to anesthesia providers.**Keywords:** Airway Management • Ambulatory Surgical Procedures • Mandibulofacial Dysostosis, Treacher Collins Type, Autosomal RecessiveFull-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/931974>

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Background

Treacher Collins syndrome is a condition that impairs bone and soft tissue development in the face. It is an autosomal dominant disorder with an incidence estimated to be 1 in 50 000 live births [1,2]. Common features include micrognathia and underdevelopment of the zygomatic arch without developmental delay [1-3]. Micrognathia and obstruction of the hypopharynx can result in difficulty breathing, to the point of becoming a grave concern. Patients with Treacher Collins syndrome often require multiple surgeries to correct facial abnormalities or underdevelopment of facial structures.

The primary anesthetic concern in patients with Treacher Collins syndrome is a difficult airway. Mask ventilation and visualization for endotracheal intubation can be challenging. The Cormack-Lehane grade reportedly worsens as patients with Treacher Collins syndrome age [1,4]. A thorough literature review helps establish standards of safety for airway management. A case review of anesthesia use in 240 of these patients at various institutions showed that 40% of them required a technique other than direct laryngoscopy to secure the endotracheal tube [4]. Placement of a supraglottic airway (SGA) device has been shown to be effective, with zero incidence of inability to ventilate [4]. Video-assisted laryngoscopy (GlideScope™ and fiberoptic) has been shown to consistently improve Cormack-Lehane grade [5,6]. SGA devices have been documented as effective conduits for fiberoptic placement as a possible first step in airway management [7-11]. The authors of 2 reports have concluded that an SGA device was “a good choice of airway when endotracheal intubation is not needed” [4,12]. In our case, we further showed that while appropriate preparation and caution is important for the management of patients with Treacher Collins syndrome, the use of an SGA device can provide safe and effective airway management. The patient’s family provided written consent to publish the present case report and personal details have been removed from it.

Case Report

Our patient was a 15-year-old boy with Treacher Collins syndrome who presented to our Comprehensive Pre-anesthesia Assessment Center for evaluation before undergoing bilateral autologous midface fat grafting. His appropriateness for treatment as a pediatric case in our ambulatory surgery center was assessed and a request also was made for him to be treated as a regular surgical case. During the evaluation, the patient’s experience with anesthesia and airway notes from Nationwide Children’s Hospital were reviewed to gauge the severity of his airway abnormalities and possible history of difficult airway. The anesthesiologists also spoke with the preoperative



Figure 1. Video and direct laryngoscopy performed for educational purposes show that the patient’s airway is Cormack-Lehane grade 1.



Figure 2. A size 3 i-Gel™ supraglottic airway device with appropriate seal for mechanical ventilation during anesthesia.

assessment nurse to coordinate gathering of photos of the patient from the primary team.

Photographs from the ear nose and throat (ENT) plastic surgeon showed that the patient had a mild ophthalmologic/otologic phenotype, although there were no images of his airway. A literature review showed that more than 40% of patients with Treacher Collins syndrome required a technique other than direct laryngoscopy to secure an endotracheal tube. An SGA device was found to be an effective airway tool, with no incidence of inability to ventilate. After discussion with the ENT surgeon, several anesthesiologists, and the medical director of our outpatient surgery center, the patient was deemed an appropriate candidate for surgery with general anesthesia. A plan was made to use an SGA device, with a fiberoptic scope and video laryngoscopy available in the room before induction. The surgeon and a senior resident/fellow also were prepared to create a surgical airway in the event of that the SGA device placement was a failure.

On the day of surgery, the patient had a grade II Mallampati score, with an oral opening and thyromental distance >3 fingerbreadths. He was given 2 mg of i.v. midazolam for anxiety. For teaching purposes, combined i.v./inhaled induction was performed with administration of propofol and nitrous oxide/sevoflurane. Mask ventilation was easy without airway adjuvants. Video and direct laryngoscopy were performed for documentation and educational purposes showing grade 1 views (**Figure 1**). A size 3 i-Gel™ SGA device was placed and an appropriate seal was achieved with no complications (**Figure 2**). The remainder of the patient's course was uncomplicated; the SGA device was safely removed on emergence and there were no adverse events after the surgery.

Discussion

Managing patients with Treacher Collins syndrome requires a comprehensive preoperative evaluation and anesthetic plan [13]. Integration of surgical, nursing, and anesthesia services determines the patient's appropriateness for surgery at an outpatient surgery center. Immediate availability of different airway devices, along with effective communication among the anesthesia team, surgeons, and intraoperative staff, mitigates the risk of a difficult airway. Video laryngoscopy has been shown to significantly improve airway visualization in what would have previously been considered an extremely difficult airway for traditional laryngoscopy techniques [14]. There is some evidence that use of an SGA device is not always successful in patients with Treacher Collins syndrome. In 2 of 7 patients with the disorder syndrome in a small case series, there was potential for epiglottic obstruction when placing the device, upon visualization with a fiberoptic bronchoscope [15]. Interestingly, in this study, a specific type of SGA device – a laryngeal mask airway (LMA) – was used. The paucity of literature regarding newer technologies in this area was evident from our literature search; practitioners do not have a strong body of work with which to guide their clinical decision-making [16]. Given the proliferation of improved airway techniques, it would be prudent to further investigate whether it is viable to treat these classically challenging patients in ambulatory settings, especially if there is evidence of potentially better controlling the airway, depending on the brand of device, such as an LMA versus i-gel or the type of video laryngoscope.

The state of healthcare following the COVID-19 pandemic underscores the importance of expanding ambulatory surgical case volumes to reach at-risk populations. Ambulatory surgical centers potentially have lower patient density and may be associated with less risk of exposure to individuals who are critically ill with COVID-19. All of these factors would be

advantageous to patients with Treacher Collins syndrome, who may have baseline respiratory abnormalities, as long as surgery can be performed safely in the ambulatory setting without significant perioperative airway complications.

Conclusions

Patients with Treacher Collins syndrome who present for surgery have potentially difficult airways, especially when endotracheal intubation techniques are used with traditional direct laryngoscopy. Also, demand is growing for performing surgery in outpatient surgery centers to decrease healthcare costs and limit patient exposures during times such as the COVID-19 pandemic. A potentially difficult airway raises questions about the safety of performing surgical cases in ambulatory settings that may lack additional staff or resources more commonly available in hospitals with inpatient and/or critical care management capabilities. However, a difficult airway is not an absolute contraindication to performing surgery on a patient in an ambulatory surgery center. Technological advancements such as improved video laryngoscopy and a fiberoptic scope for intubation can reduce the perioperative risks associated with these cases. In addition to alternative means of controlling the airway in a patient with Treacher Collins syndrome, such as with fiberoptic intubation, there is evidence to suggest that using an SGA device is appropriate in these individuals, including in the ambulatory setting. A thorough literature review of airway management techniques for a potentially difficult airway, along with comprehensive perioperative screening, can facilitate medical optimization of a patient to mitigate risks. Having a preoperative screening center, a process that allows ample time to properly assess patients and perioperative risks, and communicating with all necessary parties including the patient is paramount. Proper perioperative planning, screening, and examination of patients with Treacher Collins syndrome must be implemented before final decision-making. Given the rareness of this disease and the small volume of literature about it, case reports such as the present one must continue to be published and read so that perioperative patient safety can be improved.

Conflict of Interest

None declared.

Declaration of Figures Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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