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Goldenhar syndrome: An ephialte for the anesthesiologist – What we saw was just the tip of the iceberg

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ABSTRACT

Goldenhar syndrome, also called oculoauriculovertebral dysplasia, is characterized by craniofacial, central nervous system, cardiac, renal, and vertebral anomalies. It occurs due to the faulty unilateral development of vertebral arches. Patients presenting for surgery are usually in the pediatric age group. General anesthesia is the preferred mode of anesthesia considering the type of surgery, age of the patient, and vertebral anomaly in this patient group. The first and foremost consideration while anesthetizing these patients is the anticipated difficult airway. Frequently encountered airway abnormalities in these patients are micrognathia, retrognathia, mandibular dysplasia, craniovertebral anomalies leading to limited neck flexion and cervical spine instability, decreased thyromental distance, fascial asymmetry, cleft lip and palate, high arched palate, dental abnormalities, and temporomandibular joint abnormalities. Difficult airways often come disguised as syndromes, and thorough pre-operative evaluation along with preparation for the worst scenarios is a must. We have a myriad of alternative airway management equipment at our disposal. Adequate knowledge is a must for their best use.

Key words: Goldenhar syndrome; Difficult airway; Anticipated

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INTRODUCTION

Goldenhar syndrome is a congenital disorder with an incidence of 1:35,000–1:56,000 and a male-to-female ratio of 3:2.[1] It can occur sporadically or be inherited in an autosomal dominant or recessive pattern. It is also called oculoauriculovertebral dysplasia and is characterized by craniofacial, central nervous system, cardiac, renal, and vertebral anomalies. It occurs due to the faulty unilateral development of vertebral arches.[2] It can be associated with vater or charge syndrome with Treacher-Collins syndrome in its differential diagnosis. Here, we present a case of 12-year-old female with complete external auditory canal atresia posted for reconstruction.

CASE REPORT

A 12-year-old female child with left-side hemifacial deformity (Figure 1), a known case of Goldenhar syndrome, was posted with us for external auditory canal reconstruction (Figure 2). She had a history of bilateral temporomandibular joint ankylosis release at the age of 8 years and a history of surgery for rib cartilage graft harvest at the age of 10 years. On airway examination, she had adequate mouth opening (3 fingers), malampatti grade 3, reduced thyromental distance, receding mandible, and limited neck flexion. Her developmental milestones were normal, with no history of cardiovascular or respiratory abnormalities. All her investigations, including the X-ray soft tissue neck, were normal.

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Consent was taken from the patient's attendants, explaining the risks pertaining to a difficult airway. A difficult airway cart with a needle cricothyroidotomy and tracheostomy set was kept ready, and the otorhinolaryngologist was informed. The patient was shifted to the operation theater. ASA standard monitors for ECG, SpO_2, and NIBP were attached. In view of adequate mouth opening, we planned for direct laryngoscopic intubation with a flexible fibroscope standby. The patient was pre-oxygenated and pre-medicated with injection fentanyl 2 mcg/kg IV. Anesthesia was induced with injection propofol 2 mg/kg IV. After confirming adequate bag-mask ventilation, the short-acting muscle relaxant injection succinylcholine 2 mg/kg IV was given. Direct laryngoscopy showed Cormack-Lehane grade 4. Therefore, the decision to insert iGel 3 was taken. Adequate ventilation was confirmed with $ETCO_2$ tracing. A fiber-optic bronchoscope was used with iGel *in situ,* and the number 6 endotracheal tube was inserted under fiber-optic vision (Figure 3). Anesthesia was maintained with oxygen, nitrous oxide, and sevoflurane, and surgery went uneventfully. The patient was extubated at the end of surgery and shifted to the post-anesthesia care unit. No airway complication in the form of laryngospasm, hoarseness of voice, or blood on the supraglottic device was encountered.

DISCUSSION

Patients with Goldenhar syndrome have a wide range of manifestations, such as pre-auricular skin tags, temporomandibular joint ankylosis, microtia, cleft lip, cleft palate, epi-bulbar dermoid, eye and ear abnormalities, and craniofacial and vertebral anomalies.[3] Many of them require repeated surgical corrections during the initial developmental years of life. Patients presenting for surgery are usually in the pediatric age group. General anesthesia is the preferred mode of anesthesia considering the type of surgery, age of the patient, and vertebral anomaly in this patient group.

Airway management: The first and foremost consideration while anesthetizing these patients is the anticipated difficult airway. Frequently encountered airway abnormalities in these patients are micrognathia, retrognathia, mandibular dysplasia, craniovertebral anomalies leading to limited neck flexion and cervical spine instability, decreased thyromental distance, fascial asymmetry, cleft lip and palate, high-arched palate, dental abnormalities, and temporomandibular joint abnormalities. All these make them candidates for an anticipated difficult intubation. The incidence of difficult intubation is 30–70% in this patient group.

Associated abnormalities: Cardiovascular abnormalities are found in 5–58% of these patients. The tetralogy of fallot, atrial and ventricular septal defects, situs inversus, and coarctation of the aorta are often found in these patients.[4] A thorough pre-operative examination is essential to rule them out pre-operatively.

Important points in history: The history of obstructed sleep apnea is to be taken pre-operatively because of its common association. Difficult mask ventilation is anticipated in these patients.

The presence of vertebral anomalies such as cervical spine instability and scoliosis warrants special attention. Manual in-line stabilization during intubation is recommended in all Goldenhar syndrome patients.[5] Scoliotic spine disease leads to restrictive lung disease. Thorough neurologic assessment should be undertaken pre-operatively, and spine imaging should be considered in doubtful cases.

In our case, the mouth opening was adequate for Malampatti grade 3; therefore, our plan was endotracheal

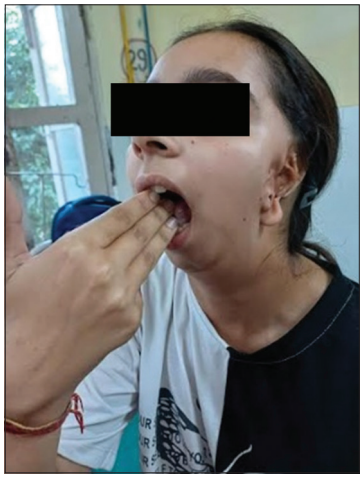

Figure 1: Patient had adequate mouth opening (3 fingers) receding chin and microtia. The narrowing of pharyngeal space and crowing of dentition make laryngoscopy impossible

Figure 2: Microtia correction

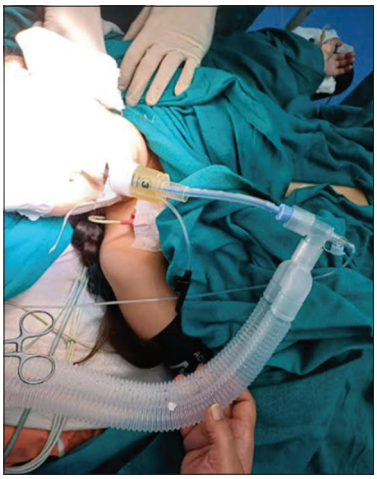

Figure 3: Intubation through iGel (smaller tube size 6 inserted, iGel size is 3)

intubation with direct laryngoscopy, and a fiberscope was kept on standby. Mask ventilation was adequate. On direct laryngoscopy, Cormack Lehane was in grade 4. Hence, iGel was inserted before moving on to the next plan. iGel displacement was a concern due to the surgical requirement of turning the face to one side; therefore, fiberoptic intubation through iGel was done and iGel was kept *in situ*. Similar to our case, Kim et al., did fiberoptic intubation through iGel in a patient with Goldenhar syndrome.[6] However, they removed iGel after intubation. Due to the increased risk of endotracheal tube displacement by iGel removal, we preferred to keep it *in situ*. Supraglottic airway devices are excellent rescue devices in difficult airway scenarios, but a definitive airway is necessary in cases of orofacial surgery. Supraglottic airway devices have a tenacious role in difficult airways.

Sun et al., did oral fiberoptic intubation using a McGrath video laryngoscope for tongue displacement in a patient with Goldenhar syndrome.[7] The purpose of tongue displacement is to create space for intubation because of oropharyngeal space constriction due to mandibular hypoplasia. Similarly, Iravani and Wald held the tongue with surgical gauze, and fiberoptic intubation was done.[8] Xing et al., encountered a case of difficult intubation in Goldenhar syndrome due to subglottic stenosis and suggested the use of a computed tomography neck to evaluate the airway pre-operatively.[9]

Appropriate preparation with an upright team can overcome a tedious situation, such as in the present case.

CONCLUSION

Difficult airways often come disguised as syndromes, and thorough pre-operative evaluation along with preparation for the worst scenarios is a must. We have a myriad of alternative airway management equipment at our disposal. Adequate knowledge is a must for their best use.

Consent for publication of the case report: A written informed consent was taken from the patient for the publication of this case report. The patient was assured that the initials would not be disclosed, though anonymity cannot be guaranteed.

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