Medical and Research Publications
International Open Access

Case Report **Journal of MAR Case Reports (Volume 6 Issue 5)**

Goldenhar Syndrome, Difficult Airway – A Case Report

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Received Date: December 31, 2022

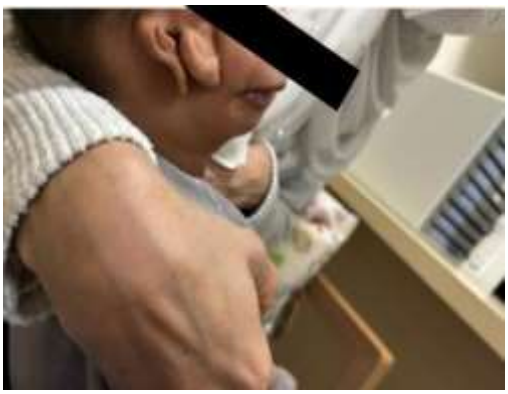
Published Date: January 10, 2023

Abstract

The Goldenhar syndrome is a rare condition which was described initially in the early 1950s by Maurice Goldenhar ***[1]****. It is characterized by a combination of congenital anomalies, mostly unilateral, including ocular (epibulbar dermoids. lipodermoids)* ***[4]*** ***[6]****, eyelid coloboma, aural (preauricular tags, anotia, microtia and hearing loss), hypoplastic facial bones, and micrognathia* ***[9]*** *and is usually associated with difficult airway. In 1963, Gorlin et al suggested the name, oculo- auriculo-vertebral (OAV) dysplasia due to its association with vertebral anomalies as part of this syndrome* ***[2]*** ***[3]****. These includes scoliosis, hemivertebrae and cervical fusion* ***[9]****. The etiology of this rare disease has not been fully understood, as it has shown itself to be variable genetically and to be caused by abnormal development of first two branchial arches* ***[9]*** ***[11]****. Anesthetists deal with the challenges of congenital anomalies including cardiac malformations (Tetralogy of Fallot, ventricular septal defects) and cleft lip and palate* ***[5]*** ***[7]*** ***[8]*** *etc. Airway management during anesthesia needs careful attention and, as most of the patients are of pediatric group, awake intubation is out of scope. We are reporting a case of Goldenhar syndrome with difficult airway, managed by using C-MAC videolaryngoscope.*

Case Description

A one-year-old boy, 6.3 kg weight with Goldenhar syndrome was brought in our hospital for ABR (Auditory brainstem response), bilateral myringotomy and Grommet insertion. Child's medical record revealed Right hydronephrosis, inability to swallow and was on NG feed. The baby was born prematurely at 36 weeks of gestation. He was admitted to neonatal intensive care unit (NICU) due to respiratory distress having stridor. He remained on high flow oxygen by nasal cannula for 18 days. After which, stridor improved and baby was discharged home. He did not have any history of allergies or surgery.



On his preoperative anesthesia clinic visit, we found that there was gross facial asymmetry. The Right side of the face was malformed including hypoplastic mandible and cheek bones, malformed nose, right auricular appendages and Right anopthalmia. Mallampati scale was 4 with normal neck mobility. Other routine laboratory tests, echocardiography and x-ray chest were within normal limits.

Arrangements were made for a difficult airway including C-MAC videolaryngoscope, pediatric fiberoptic bronchoscope, bougie, size 1 and 1.5 laryngeal mask airways (LMA), endotracheal tubes of several sizes with stylets. Preoperative glycopyrrolate was given half an hour before bringing the child to operation room. After attaching standard monitors, inhalational induction was done with

incremental percentages of sevoflurane in oxygen. Once the child was deep enough, laryngoscopy with C-MAC done. Initially, vocal cords were not visible but by manual laryngeal manipulation, arytenoids and posterior end of vocal cords were seen. Once confirmation of the ability of intubation, we gave intravenous fentanyl and placed LMA size 1. During the procedure, LMA worked well and the child was kept on pressure support ventilation (PSV). After around 40 minutes of the procedure, the child was awakened. The baby tolerated the procedure and anesthesia well and vitals remained stable. His postoperative stay in postoperative care unit (PACU) and hospital was eventless. The child was discharged home on the same day. Later, the ENT consultant advised him bilateral cochlear implant.

We discussed this patient in departmental meeting and all the colleagues were informed about the patient as well as other comorbidities associated with Goldenhar syndrome were discussed. The consensus was that the difficult airway management starts from good preoperative anesthesia assessment. It is a core of any difficult airway followed by the application of the difficult airway algorithm. Team work is also fundamental in airway management. It was suggested that more than one anesthetist can also be helpful.

Patient's parents were informed about the possibility of difficult airway from the time of preoperative anesthesia clinic visit. After the procedure, they were briefed again for any future anesthesia. The airway assessment and management were highlighted in hospital's medical record as well for future reference.

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