

# Our anesthesia experience in a patient with goldenhar-gorlin syndrome

## Abstract

Childrens with congenital anomalies affecting the airway can pose a significant challenge for the anesthesia management. In Goldenhar-Gorlin syndrome, anomalies that may lead to difficulty in intubation anomaly may occur. Difficult or impossible laryngoscopy has been described in Goldenhar- Gorlin syndrome patients. The aim of this case report is to attract attention to this syndrome which fulfills many criteria for difficulty of intubation.

**Keywords:** goldenhar syndrome, airway management, difficult intubation

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**Ozkan Onal, Emine Cepni Kutahya, Oguzhan Arun, Ates Duman**

Department of Anesthesia and Intensive Care, Selçuk University Medical Faculty, Turkey

**Correspondence:** Ozkan Onal, Department of Anesthesiology and Reanimation, Selçuk University Faculty of Medicine, Konya, Turkey, Tel +905059052252, Email drozkanonal@selcuk.edu.tr

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## Introduction

Goldenhar-Gorlin syndrome was defined by Goldenhar in 1952 and redefined by Gorlin et al.<sup>1</sup> in 1963 as auriculo-vertebral syndrome. Goldenhar syndrome is a rare oculo-auriculo-vertebral syndrome occurring at the stage of blastogenesis resulting from 1 and 2 branchial arch anomaly. It occurs every 1/3500-1/5600 births. Male/female ratio is: 3/2.<sup>2</sup> Goldenhar-Gorlin syndrome is characterized by ear anomalies, hemifacial microsomia, cardiac anomalies and vertebral column anomalies. In this syndrome, anomalies that may lead to difficulty in intubation such as facial asymmetry, large forehead, micrognathia, cleft palate, hemifacial hypoplasia, malocclusion and vertebral anomaly may occur. The aim of this case report is to attract attention to this syndrome which fulfills many criteria for difficulty of intubation.<sup>3,4</sup>

## Case

A 9 year old patient at the weight of 30kg and who was diagnosed with Goldenhar-Gorlin syndrome was planned to undergo meatoplasty operation by otorhinolaryngology department. In the anesthesia evaluation prior to operation, it was learned that he had no chronic disease and known allergy other than the syndrome. In preoperative evaluation, the following features of the syndrome were present: facial symmetry, preauricular tag, epibulbar dermoid, right maxillofacial hypoplasia and right mandibular hypoplasia, torticollis, scoliosis, membranous ventricular septal defect anomalies which were reported to have closed previously and gastroesophageal reflux. Head and neck movements were slightly restricted due to torticollis and overall Wilson risk score was 2 (weight: 0, head-neck movements: 1, jaw movement: 0, jaw structure: 1, teeth structure: 0). In the anesthesia evaluation made before operation, it was learned that he underwent operation when he was 7 months old due to urachus cysts and at the age of 2 due to adenotonsillectomy. No information could be obtained regarding anesthesia techniques used during these operations. Laboratory findings and lung graphy findings were normal except for scoliotic minimal curve in thoracic vertebrae. As the patient fulfilled the criteria of difficult intubation, prior to operation airways, ventilation masks, endotracheal tubes, Miller and Macintosh blades appropriate to the child's age were prepared. Patient was transferred to operating theater and routine monitorization was performed. The patient was agitated, and induction was initiated with sevofluran

and  $N_2O$  and vascular access was established. 10 mg Rocuronium (Esmeron, Organone, Hollande) was administered because of its fast effect. After ventilation with mask, he was intubated without any problem with gum intubation catheter (Frova, Cook, USA). Anesthesia was maintained with the mixture of 60%  $N_2O$  and 2.5% sevofluran. Following the operation lasting one hour, spontaneous respiration returned and after 0.3 mg atropin and 1.2mg neostigmine were administered, patient was extubated and sent to clinic without any complications.

## Discussion

In patients with Goldenhar-Gorlin syndrome, the presence of craniofacial and vertebral anomalies increases the risk of difficult airway. In addition, whether operation is carried out under emergency or elective conditions is important for securing airway.<sup>5</sup> In the evaluation made in the patients with Goldenhar-Gorlin syndrome, scoliosis, micrognathia and restriction of extension are risk factors for the probability of difficulty of intubation. Therefore one must be ready for nasal laryngeal mask, fiber optic intubation, and if necessary tracheostomy. These patients may also experience ventilation problems due to decrease in functional and residual capacity and total lung capacity accompanied by hemifacial hypoplasia. Moreover, a patient with Goldenhar-Gorlin syndrome may also have obstructive sleep apnea commonly. Altıntaş F & Cakmakkaya OS.<sup>6</sup> decided to intubate a 4 year old patient with Goldenhar-Gorlin syndrome who will undergo ectropion surgery without muscle relaxants and administered propofol (Propofol, Fresenius, Turkey) (2.0mg/kg) and remifentanyl (Ultiva, Glaxo SmithKline, Turkey) (1µgr/kg) as anesthetic. They did not encounter any problems in anesthesia induction when they were not successful at first attempt with Macintosh 2 blade, they passed to Miller 2 blade and attempted intubation with 4.5mm tracheal tube. They did not succeed at first attempt, but tube number was made smaller (4mm) and intubation attempt was repeated with the help of intubation stilette and with success this time. In the maintenance of anesthesia 1% sevofluran was used with mixture of %50  $O_2/N_2O$  and anesthesia was completed without complication, not using any muscle relaxants. In the present study, propofol was not preferred as it is still controversial in pediatric age group before the age of 3, and for anesthesia induction, midazolam (0.1mg/kg) and fentanyl (1µgr/kg) were used. Kabalak et al.<sup>7</sup> attempted blind intubation in a 6 month year old patient planned to undergo operation for congenital

hip dislocation using muscle relaxants and were successful at second attempt via an intubation tube sent as a guide to oesophagus. Jones et al.<sup>8</sup> used laryngeal mask for securing airway in all of the 43 patients with Goldenhar-Gorlin syndrome between the ages of 23 months - 14years who were anesthetized for the implanting of hearing device and emphasized that laryngeal mask provides easy, safe and rapid anesthesia without leading to trauma. Sukhupragam W & Rosenblatt WH<sup>9</sup> preoxygenized a 10year old patient, who was planned to undergo keratoplasty and predicted to have difficulty in airway and then deepened the anesthesia with sevofluran induction the concentration of which they increased gradually. In order to facilitate airway manipulation, flexible laryngeal mask was placed readily using 50mg propofol. For maintenance of anesthesia, they used 30% O<sub>2</sub> / 70% N<sub>2</sub>O mixture with 2% sevofluran and did not employ muscle relaxants in any stage of anesthesia and as analgesic they used overall 30 µgr fentanyl with intervals. Alternative airway devices developed in parallel to the advances in technology facilitated the job of anesthetists, increasing the safety of both the patients and the anesthetist. Milne et al.<sup>10</sup> reported that in a 9 year old girl with Goldenhar-Gorlin syndrome who will be submitted to general anesthesia for ear debridement and tooth extraction, when they were not successful in direct laryngoscopy which they attempted without muscle relaxants by using 8% sevofluran induction and intermittent propofol (2mg/kg), they obtained images at adequate quality after a few attempts with a video laryngoscopy method called glidescope and managed to intubate the patient. Özlü et al.<sup>11</sup> reported that, in a 6 year old child with Goldenhar-Gorlin syndrome planned to undergo operation for posterior fossa decompression and occipitocervical fusion after premedication with, midazolam (7mg), nasal topical anesthetics were administered and patient was intubated at the first attempt with fiberoptic bronchoscopy. Maintenance of anesthesia was made with desfluran 50%/50% O<sub>2</sub>/air mixture using muscular relaxant (0.1mg/kg vecuronium). Although awake intubation is gold standard in patients who are estimated to have airway difficulty, this is not possible in pediatric patients as it may lead to airway anomalies.<sup>9</sup> In the present patient predicted to have airway difficulty, mask ventilation and intubation was carried out successfully at first attempt, which relieved us and obviated the need for alternative airway techniques.

## Conclusion

As the intubation is highly likely to be difficult in such patients, a thorough preoperative evaluation is required. It is our suggestion that adequate monitorisation should be carried out, anesthesia technique and equipment and technique should be determined in advance and necessary preparations is made for successful intubation and intubation stile can be used for difficult intubation.

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## Conflicts of interest

The authors declare that there are no conflicts of interest.

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