Airway Management In A Patient With Goldenhar Syndrome Using Classic Laryngeal Mask Airway: A Case Report

Maureen Thea C. Estrada, MD
Vanessa G. Jamora, MD, FPBA
Josefina T. Policarpio, MD, FPBA

Department of Anesthesiology
University of Santo Tomas Hospital

ABSTRACT

Goldenhar Syndrome is an embryonic developmental anomaly involving the first and second arches resulting in a wide spectrum of birth defects including ocular, auricular, facial, cranial, vertebral and cardiac anomalies. Among the various deformities associated with the syndrome, hemifacial microsomia, maxillo-mandibulo-facial hypoplasia, and neck bones fusion are anesthesiologist's concern because these features are precursors to difficult airway management, particularly so that its corrective surgical procedures are to be done under general anesthesia. For the excision of the right ocular dermoid with plastic-reconstructive skin grafting of this 9 years old female patient, we were able to prevail over this dilemma by using a right fitting face mask and classic flexible LMA while she was breathing spontaneously under Sevoflurane general anesthesia. Early recognition and keen knowledge of the disorder and proper preparation for its anticipated anesthetic implications are the foundation of successful peri-operative management of patients with Goldenhar Syndrome.

Keywords: *Goldenhar Syndrome, Oculo-Auriculo-Vertebral Spectrum, Classic LMA general anesthesia*

INTRODUCTION

Oculo-Auriculo-Vertebral Spectrum (OAVS) refers to three rare congenital anomalies that although presenting as poorly understood disorders with diverse manifestations and severity, are inter related to each other. Goldenhar Syndrome being the most severe form has most of the manifestations of Oculo-Auriculo-Vertebral disorder (OAVD): the mildest form of disorder, and of Hemifacial Microsomia (HFM): the intermediate form. Reported occurrence of OAVS is estimated to range from one in 3,000 to 5,000 live births up to one in 25,000 to 40,000 live births; male:female ratio of 3:2.[1]

As first described in 1952 by the Swiss ophthalmologist, Maurice Goldenhar,[2] Goldenhar Syndrome involves complex developmental disorders of the face, ears, eyes, spine, etc., manifesting with varying severity in each patient: as facial asymmetry due to hypoplasias of the facial muscle, of the maxillary and mandibular bones and the zygomatic arches (hemifacial microsomia); micrognathia; external ear malformations and hearing loss; ocular dermoids or lipodermoids. Other oral cavity anomalies may include a high-arch or cleft palate and abnormalities of the tongue.[3]

Children with Goldenhar syndrome are known to present with airway management challenges. Difficult intubation may be expected due to mandibular hypoplasia and fused underdeveloped neck bones, among others.[4]

It is our objective to document a case of Goldenhar syndrome; presenting the airway and other anatomical conundrum that accompanies it, and to propose anesthetic approach to patients with this syndrome.

CASE PRESENTATION

This is a case of a 9 year-old, 20-kg, 123cm female patient who was admitted for excision of an ocular dermoid with membrane graft of the right eye.

She was observed to be hard-of-hearing with difficulty in speaking. Other significant findings were: hemifacial microsomia, hypoplasia of the malar bones and mandible, marked micrognathia, microtia and right ocular dermoid.

Figure 1 Facial assymmetry

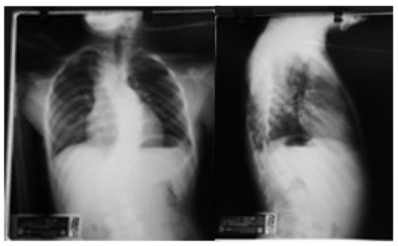

Figure 2 Moderate Levoscoliosis

Her head was tilted to the left, right shoulder higher than the left, the neck had limited movement, the cervical vertebrae are fused, the spine was S shaped with right 'rib hump'; chest XRay: moderate scoliosis.

At the preanesthesia room, she was premedicated with midazolam 0.05mg/kg, ketamine 1mg/kg and atropine 0.01mg/kg intravenously. Once she was sedated, she was brought to the OR suite.

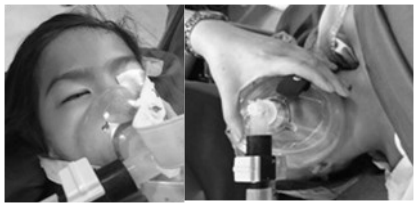

Figure 3 anesthesia face mask fit

We had difficulty fitting the size 2 tear drop shaped standard anesthesia face mask to the right side of her face. The size 2 round shaped flexible anesthesia face mask made a perfect fit without gas leakage and without upper airway obstruction. Induction was carried out with sevoflurane 2vol% then propofol 2mg/kg/IV prior to insertion of size 2.5 classic LMA (undeflated). After inflation of 5 cc of air on the

balloon, no leak was observed; the breath sounds were heard distinctly without air turbulence and the chest moved smoothly while the patient was breathing spontaneously. Capnography was 30 mm Hg.

Anesthesia was maintained with sevoflurane at 2vol% on spontaneous-manually assisted respiration. During emergence, the patient was allowed to self-expel the LMA without coughing or retching.

DISCUSSION

Goldenhar syndrome is a sporadic congenital anomaly of unknown etiology, usually presenting with hypoplasia of the facial muscle, of the maxillary and mandibular bones, of the zygomatic arch and underdeveloped vertebral bones. In majority of cases the deformity is unilateral with 3:2 right-side to left-side prevalence. [1]

Like in 77% of Goldenhar syndrome, this female child had hemifacial microsomia with gross facial asymmetry, severe micrognathia, difficulty in speaking and microtia with deafness. The epibulbar dermoid that was to be removed is also seen in 39% of cases.

Several reports of difficult airway management in pediatric patients with Goldenhar syndrome had been cited in the literature.[3,4,6,8] Compounding the picture was the presence of vertebral anomaly that is exhibited in 70% of this syndrome. Fused neck bones limiting the neck movement can be a major contributor to improper exposure of the laryngeal inlet; this is notwithstanding the possibility that the mandibular bone hypoplasia can alter the anatomy of the hypopharynx. This could have been the underlying cause of unsuccessful intubation reported by Shukry et al[8] and Sugino et al[9] both in 2005: the former resorted to using a laryngeal mask while the latter turned to using a video laryngoscope for intubation. There are even documented reports of patients with Goldenhar syndrome whose operations were deferred because they could not be intubated. [3,4]

The anesthetic technique however, can be adapted to the type and site of the surgical procedure to be done. For the excision of an ocular dermoid with skin grafting, although there is no dire need for endotracheal intubation, this patient still needed adequate depth of general anesthesia. For the same kind of operation Sukhupragarn in 2008 used fexible LMA in a 10-year old patient with Goldenhar syndrome.[11] There were likewise previous reports showing successful use of the LMA in children with severe micrognathia such as that associated with Treacher Collins syndrome and Pierre Robin syndrome.[7]

Even at first glance during inspection, we were able to predict glitches in airway management. True enough we could not even fit the conventional tear shaped anesthesia face mask on the affected side for lack of facial muscle, maxillo-mandibular bones and zygomatic arch to hold and support the soft tissues of the face. We were able to overcome this predicament by shifting to a round shaped face mask; but had the asymmetry and all the above facial disconfigurations been placed on the left side, we could have encountered some challenge in placing her head on the chin lift position because we had nowhere to anchor our fingers on the scenario of left mandibular hypoplasia. The positioning of the head would likewise be more difficult had the cervical vertebra anomaly tilted the head and neck to the right. Although we exercised great caution by not extending her head so as not to compromise the cervical vertebral pathology we could have not guaranteed not doing this maneuver had we opted on intubating this patient.

For the insertion of the LMA, an adequate level of anesthesia must be attained to prevent laryngospasm, and likewise to achieve the best fit on the hypopharynx. In the study of Dr. V. Priya et al in 2002, excellent conditions for LMA insertion were obtained in significantly greater number of patients with Propofol induction than with Sevoflurane gas induction. [19] But 'single breath' Sevoflurane induction as done by these authors is not that feasible in children, while the 2.5 mg/kg Propofol dose may be large enough to precipitate apnea which we have to avoid in patients with probable difficult airway.

To provide depth of anesthesia that could adequately suppress the laryngeal reflexes, we opted on letting this patient breath 2% Sevoflurane; then

once assured that she could be well ventilated, we administered Propofol at 2 mgs/kg.

With this approach, we were also able to circumvent the spasticity that Sevoflurane may invoke on the normal facial muscle and jaw:[19] a situation that may further aggravate the airway's anatomical deformity. Moreover, Propofol has been documented to depress the laryngeal reflexes. [19]

As per recommendation for difficult airway, we did not use a muscle relaxant.

Another point of deliberation we had was that in the presence of right facial muscle and mandibular hypoplasia, the pharyngeal structures might have been pulled to the left; in which case, the LMA cuff might not sit well in the hypopharynx to adequately cover the laryngeal inlet.[20] Ventilation would then become less effective and most importantly the risk of intraoperative aspiration would increase.

Nevertheless, we put emphasis on choosing the right type and optimal size of the LMA and inflating the cuff with the minimum amount of air that can create just enough airtight seal. This way, the Classic LMA we inserted was able to adapt to this patient's distinctive upper airway anatomy. This flexibility of Classic LMA was also demonstrated by Florendo et al. (2014) in their pediatric patient with Hallerman Streiff syndrome.[21] These experiences just confirmed earlier observations that although pediatric Classic LMA is a 'scaled-down version' of the adult model, it was used successfully albeit differences seen between pediatric and adult airways. [12] Additionally, we were able to bend the Classic LMA to give more elbow room for the surgeon and his bulky microscope.

CONCLUSIONS

Hemifacial microsomia, gross facial asymmetry, fused neck bones are just part of the conundrum of anatomic defects in Goldenhar syndrome but these would suffice to push the alarm button for difficult airway management.

Preoperative evaluation therefore of children with Goldenhar syndrome begins with evaluation of the upper airway, with a high suspicion of anticipated difficulty, and formulation of a plan for airway management.

Anaesthetic management has to be tailored to the clinical situation and surgical procedures to be done. With a well fitting round shaped flexible anesthesia facemask and optimal depth of anesthesia with 2 volume % Sevoflurane and Propofol 2mgs/kg, we were able to insert a size 2.5 Classic LMA that sat snugly in this patient's hypopharynx; which stayed well all throughout the 1 ½ hours eye surgery while the patient was breathing spontaneously.

REFERENCES

1. Kokavec, R. Goldenhar Syndrome With Various Clinical Manifestations. Craniofacial Journal. 2006; 43 (5):628-634.
2. Ansari, S. et al. Goldenhar syndrome presenting as limbal dermoid cyst: A case report. International Journal of Case Reports and Images. 2013;4(7):384-387.
3. Sukhupragarn, W. and Rosenblatt, W. Airway management in a patient with Goldenhar syndrome: a case report. Journal of Clinical Anesthesia (2008) 20, 214–217
4. Roodneshin, F. and Agah, M. Management of Anesthesia in Goldenhar Syndrome: Case-Series Study. National Research Institute of Tuberculosis and Lung Disease. 2009;8(4):43-50.
5. Barash, P. et.al. Clinical Anesthesia 7th ed. 2013. P.762.
6. Mehta B., et.al. *Goldenhar syndrome with unusual features.* Indian Journal of Dermatology. 2008; 74(3): 254-6.
7. Ortega, U. Management of Anesthesia and the airway in a child with Goldenhar syndrome. Servicio de Anestesia y Reanimacion. 2001; 48:97.
8. Shukry M. et al. *Management of the difficult pediatric airway with Shikani Optical Stylet.* Pediatric Anesthesia. 2005; 15(4): 342-345.
9. Sugino, S., Kanaya, N., Namiki, A. *Anesthetic management in a patient with Goldenhar's syndrome using a perilaryngeal airway and a videolaryngoscope.* Masui 2005; 54(12): 1367-70.
10. Cardwell M, Walker RWM. *Management of the difficult paediatric airway*. British Journal of Anesth 2003; 3(6):167-170

11. Madan, R. et.al. *Goldenhar's Syndrome: An analysis of anesthetic management A retrospective study of seventeen cases.* Anaesthesia. 1990; 45:49-52.

12. Kim H, Park M, Kim J, Kim S, Kim H. *Appropriate laryngeal mask airway size for overweight and underweight children*. doi:10.1111

13. Rommel, N. et al. *Development of the orohypopharyngeal cavity in normal infants and young children.* The Cleft Palate–Craniofacial Journal 2003; 40: 606-11.

14. Singh, D. and Bartlett, S. *Congenital Mandibular Hypoplasia: Analysis and Classification.* The Journal of Craniofacial Surgery. 2005; 16(2): 291-300.

15. Nagler, J and Bachur, R. *Advanced airway management.* Emergency and Critical Care Medicine.2009. DOI:10.1097/MOP.0b013e32832b112c

16. Caplan,R. *American Society of Anesthesiologists Task Force on Management of the Difficult Airway.* Anesthesiology 2003; 98: 1269-1277.

17. GÖnÜl Tezcan Kele, Verda Toprak, GÜlay Ok, Nurettin LÜleci. *Volatile induction and maintenance of anesthesia using laryngeal mask airway in pediatric patients*. Journal of anesthesia 2004; 18: 51-4.

18. Pinheiro, A. et.al. *Goldenhar Syndrome- Case Report.* Brazilian Dental Journal. 2003. 14(1); 67-70.

19. Afzal, M. *Airway Management in Pediatric Anesthesia: Laryngeal Mask Airway Vs Endotracheal Tube.* 2006. 13(1); 1-3.

20. Priya, V., Deviata, J., and Dasgupta, D. *A Comparison of Propofol Versus Sevoflurane for Laryngeal Mask Airway Insertion*. Indian Journal of Anesthesia. 2002; 46 (1): 31-34.

21. Patel, B. and Bingham, R. *Laryngeal mask airway and other supraglottic airway devices in pediatric practice.* Continuing Education in Anesthesia, Critical Care and Pain. 2009; 9(1); 6-9.

22. Florendo, G. and King, L. *A Case of Hallerman Streiff Syndrome for Bilateral Under General Anesthesia.* 2014