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Anaesthetic Management in a Child with Goldenhar Syndrome

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CASE REPORT

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ABSTRACT

Goldenhar syndrome is a congenital disorder involving deformities of the face. It usually affects one side of the face only and poses significant challenges in the airway management. We herein, report an 8-year boy, known case of Goldenhar syndrome, who presented to our radiology suite for a magnetic resonance imaging (MRI) brain, followed by a computed tomography (CT) scan brain. The boy had various features of Goldenhar syndrome, e.g. cleft palate, absent right eye and ear, right mandibular hypoplasia, micrognathia, and preauricular tags. His developmental milestones were delayed. Airway evaluation showed Mallampati class II with limited movements of head and neck, which suggested possibility of difficult laryngoscopy and intubation. He had no vertebral anomalies or cardiac disease. A difficult airway continues to be a major cause of anaesthesia-related morbidity and mortality; and maintaining spontaneous breathing remains a vital technique in its management. Lack of anaesthesia-related complications with supraglottic devices encouraged us to present the advantage of utilising a laryngeal mask airway (LMA) under anaesthesia for successful management of predicted difficult airway.

Key Words: *Goldenhar syndrome. Laryngeal mask airway. MRI and CT scan.*

INTRODUCTION

During general anaesthesia, the most common causes of morbidity in the paediatric population are airway and respiratory complications.[1] Goldenhar syndrome (GS), also known as hemifacial microsomia, was first described by Maurice Goldenhar in 1952.[2] These children commonly present with difficulties in airway management. The defects are characterised by facial asymmetry and hypoplasia, hypoplastic zygomatic arch, micrognathia, external ear malformations, hearing loss, vertebral anomalies, and ocular dermoids or lipodermoids. The male to female ratio is 2:1 and its occurrence is estimated from 1 in every 3,000 to 1 in every 5,000 live births.

The cause of GS seems to be heterogeneous.[3] Tracheal intubation can be difficult. Many difficulties are relatively easy to predict prior to induction and are associated classically with craniofacial problems. In addition, the difficult airway typically becomes progressively worse with increasing age.

CASE REPORT

This patient was an 8-year, 23 kg boy, previously diagnosed with GS scheduled for MRI and CT scan. The patient had various features of GS, e.g. cleft palate, absent right eye and ear, right mandibular hypoplasia, micrognathia, and preauricular tags. His developmental milestones were delayed. Airway evaluation showed Mallampati class II with limited movements of head and neck, which suggested possibility of difficult laryngoscopy and intubation. He had no vertebral anomalies or cardiac disease and no history of any recent chest infections; therefore, no echocardiogram or chest X-ray were ordered preoperatively.

On immediate preoperative assessment, the child was otherwise healthy without any history of fever or upper respiratory tract infections. He was not premedicated. A written informed parental consent was taken after discussing risks and management of difficult airway with the parent. All preparations were made anticipating a difficult airway which included having a difficult intubation trolley having endotracheal tubes, Guedel airway and face masks of different sizes and a more senior anaesthetist on backup in case of any emergency. Before induction, standard American Society of Anaesthesiologists (ASA) monitoring electrocardiography (ECG), noninvasive blood pressure (NIBP), and oxygen saturation (SpO_2) were established. After allowing the patient to breathe 100% oxygen, induction of anaesthesia was performed with voluntary inhalation of an increasing concentration of sevoflurane (2 - 8%) via a paediatric circle system. After loss of consciousness, an intravenous catheter 24G was then placed on dorsum of right hand, and sevoflurane adjusted to 2 - 3%. A size 2.5 Laryngeal Mask Airway (LMA) was inserted after adequate jaw relaxation was attained. Inhalation agent was then switched to isoflurane and maintained with 1.2% isoflurane in 60% nitrous oxide and 40% oxygen. Propofol 10 mg/ml in a 10 ml syringe as well as atropine 0.1 mg/ml (total 10 ml) and succinylcholine 10 mg/ml (total 10 ml) were prepared and kept readily available in case of any emergency. Laryngoscope blades and different sized endo-tracheal tubes, both cuffed and un-cuffed, were also available in case of difficulty to ventilate the patient. Video-laryngoscope was also made available in case of need.

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After induction in the preoperative area, the patient was shifted to the adjacent MRI suite breathing spontaneously and all monitors reapplied. In the MRI suite, all monitors that were applied were MRI compatible including standard monitors for heart rate, pulse oximetry and SpO_2. ECG and capnography were attached on the Tesla M3 monitor. The MRI took about 45 minutes and the patient remained stable throughout the procedure. Thereafter, the patient was shifted to the CT scan room which was on the same floor in the radiology department with intermittent boluses of propofol (10 mg) while maintaining spontaneous ventilation at all times. The patient was shifted using 100% oxygen with the paediatric Bain circuit attached to oxygen cylinder. A portable monitor was also attached to the patient ensuring adequate ASA standards. Throughout the procedure, 2 anaesthetists were present with the child at all times.

The patient was kept on spontaneous breathing throughout the CT scan and remained hemodynamically stable. LMA was removed when the patient was fully awake and with return of airway reflexes. The patient's procedure and anaesthetic courses were uneventful, and he was shifted to the recovery room fully awake. His post-anaesthetic course in the recovery room was unremarkable, and he was discharged from the hospital the same day.

DISCUSSION

Patients with difficult airways are successfully managed using supraglottic devices and it can also be used as a source for intubation in difficult airways. Supraglottic airway devices have an important role in airway management of difficult airways in both adults and children.[4] However, there is limited data with respect to utilisation of supraglottic airway devices for principal airway management in children.

In this case, MRI brain showed deformed and smaller right orbit showing heterogeneous area in the anterior part which may possibly represent atrophic globe, relatively smaller right-sided maxillary antrum, absence of right sided zygomatic arch and right ramus of mandible and decreased pneumatisation of right mastoid air cells. Traditionally, the presence of ear abnormalities, which leads to the search for mandibular hypoplasia and vertebral alterations, is the main clinical feature associated with GS diagnosis.[5]

The presence of mandibular abnormalities has 100% sensitivity and 96% specificity for predicting difficult laryngoscopy. As the number of associated craniofacial anomalies of GS increases, the risk of difficult intubation also increases. The airway and anaesthetic management of the case presented was challenging as the pre-operative assessment of his airway revealed the presence of maxillary hypoplasia and limited head and neck movements with Mallampati class II.

In the paediatric population, the utilisation of an inhalational induction is favoured over intravenous induction.[6] There have been few reported cases of airway management in patients with GS. Copper and Murray-Wilson successfully used a retrograde intubation in a 5-month old infant.[7] Bakh *et al*. reported the use of fiberoptic intubation via laryngeal mask in a patient with GS.[8] Even though use of awake fibreoptic intubations in children is reported, awake intubation may at times be impossible in infants and children. Furthermore, difficult airway gadgets are not accessible in paediatric patients at times.[9]

The use of supraglottic devices has gained popularity and acceptance in the management of difficult airway. The guidelines of dealing with the difficult airway in children is to maintain spontaneous ventilation.[10] LMAs have proved to be an adequate source of maintaining the airway and at the same time proved beneficial in cases of failed intubations. Because of ease of insertion and accuracy, we have found LMAs to be quite convenient in both the adult as well as paediatric population in our practice, especially in outpatient settings such as the radiology suites.

REFERENCES

1. Aydogan M, Begec Z, Erdogan M, Yücel A, Ersoy M. Airway management using the ProSealTM laryngeal mask airway in a child with Goldenhar syndrome. *Riv Eur Sci Med Farmacol* 2012; **16**:559-61.
2. Gupta K, Subharti N, Subhartipuram N. Goldenhar syndrome: Airway and anaesthetic management: A case report. *J Pioneeri Med Sci* 2011; **1**:56-9.
3. Martelli-Júnior H, Miranda RTD, Fernandes CM, Bonan PRF, Paranaíba LMR, Graner E, *et al*. Goldenhar syndrome: Clinical features with orofacial emphasis. *J Appl Oral Sci* 2010; **18**:646-9.
4. Jagannathan N, Sequera-Ramos L, Sohn L, Wallis B, Shertzer A, Schaldenbrand K. Elective use of supraglottic airway devices for primary airway management in children with difficult airways. *Br J Anaesth* 2013; aet411.
5. Strömland K, Miller M, Sjögreen L, Johansson M, Joelsson BME, Billstedt E, *et al*. Oculo-auriculo-vertebral spectrum: Associated anomalies, functional deficits and possible developmental risk factors. *Am J Med Genet Part A* 2007; **143**: 1317-25.
6. Brooks P, Ree R, Rosen D, Ansermino M. Canadian paediatric anaesthesiologists prefer inhalational anaesthesia to manage difficult airways: A survey. *Canadian J Anesth* 2005; **52**:285-90.
7. Cooper C, Wilson AM. Retrograde intubation. *Anaesthesia* 1987; **42**:1197-200.
8. Bahk JH, Han SM, Kim SD. Management of difficult airways with a laryngeal mask airway under propofol anaesthesia. *Pediatr Anesth* 1999; **9**:163-6.
9. Milne AD, Dower AM, Hackmann T. Airway management using the paediatric GlideScope® in a child with Goldenhar syndrome and atypical plasma cholinesterase. *Pediatr Anesth* 2007; **17**:484-7.
10. Walker RW, Ellwood J. The management of difficult intubation in children. *Pediatric Anesth* 2009; **19**:77-87.