Airway Management in a Child with Goldenhar Syndrome

Yan-Hua Sun[1,2], Bo Zhu[1], Bing-Yang Ji[2], Xiu-Hua Zhang[1]

[1]Department of Anesthesiology, Peking Union Medical College Hospital, Beijing 100730, China
[2]Department of Cardiopulmonary Bypass, Fuwai Hospital, National Center for Cardiovascular Diseases, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing 100037, China

Key words: Airway; Anesthesia; Goldenhar Syndrome

Goldenhar syndrome, also known as oculo-auriculo-vertebral dysplasia, is characterized by a wide range of congenital anomalies, such as micrognathia, oral cavity malformations, and vertebral anomalies and always causes difficult airway. Awake intubation often cannot be used on children because of inadequate cooperation. In addition, children have higher rates of oxygen consumption, significantly shortening the period of apnea that can be safely tolerated. This study reported a successful orotracheal intubation using the McGrath® video laryngoscope combined with a fiberoptic bronchoscope (FOB) in a pediatric patient with Goldenhar syndrome.

A 5-year-old girl, 20 kg in weight and 114 cm in height, with Goldenhar syndrome was admitted in our hospital for electronic cochlear implantation surgery. Her medical history did not reveal any operations, chronic diseases, or known allergies. During the preoperative visit, we noted that her facial contours were abnormal, including facial asymmetry, hypoplastic mandibles, retracted lower jaw, microtia, and auricular appendages [Figure 1]. The intubation condition according to the Mallampati scale was assessed as class 4, and she displayed normal neck mobility. The preoperative laboratory parameters were all within normal limits and no abnormal findings were shown on the chest radiograph or electrocardiograph.

Several airway devices were prepared before induction including a 2# laryngeal mask airway (LMA), several sized endotracheal tubes with stylets, and the McGrath® video laryngoscope. A FOB was also available. After being transferred to the operating room, the girl was routinely monitored, including electrocardiogram, pulse oximetry, and noninvasive blood pressure. The inhalation induction was applied with 8% sevoflurane in oxygen. After venous access was established, 3 mg/kg propofol, 1 μg/kg fentanyl, and 0.5 mg/kg rocuronium were administered. The bag-valve-mask ventilation was without any difficulty during the preoxygenation and throughout the intubation process. Under the view of the McGrath® video laryngoscope, the glottis was not visible. We tried transnasal fiberoptic intubation with an endotracheal tube #6.0 twice, but both failed to visualize the vocal cords due to the patient's tongue body hypertrophy and narrow supraglottic space. Then, we attempted to use McGrath® video laryngoscope to displace the tongue and enlarge the oral cavity space. At last, a successful transoral fiberoptic intubation was made. The pulse oxygen saturation was maintained around 85–100%, and the heart rate ranged from 100 to 120 beats/min.

The patient was maintained on O_2:N_2O (1:1) and sevoflurane 2%, fentanyl was supplemented as required. The operation was completed uneventfully. The child was fully awakened and extubated in the operating room. She recovered fully without any complications and was discharged after one week.

Conscious intubation was not suitable for such a young patient; thus, we decided to intubate after induction. As it was a prospectively difficult case of glottis exposure without the difficulty of mask ventilation, we chose rocuronium for better muscle relaxation and glottis exposure, thus rejected retaining spontaneous respiration. In view of a limited oxygen reserve and the time consumed by the fiberoptic intubation, we attempted with a McGrath® video laryngoscope at first. With the McGrath® video laryngoscope, we failed to expose

Address for correspondence: Dr. Xiu-Hua Zhang, Department of Anesthesiology, Peking Union Medical College Hospital, Beijing 100730, China
E-Mail: zhangxh@pumch.cn

Received: 20-08-2017 **Edited by:** Yi Cui
How to cite this article: Sun YH, Zhu B, Ji BY, Zhang XH. Airway Management in a Child with Goldenhar Syndrome. Chin Med J 2017;130:2881-2.

Access this article online

Quick Response Code:

Website: www.cmj.org

DOI: 10.4103/0366-6999.219146

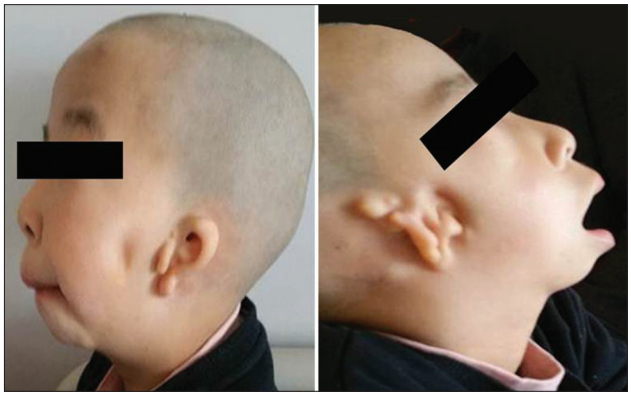

Figure 1: The patient with Goldenhar syndrome showed facial asymmetry, hypoplastic mandibles, retracted lower jaw, microtia, auricular appendages.

the glottis, even with the pressing on the neck. The failure rate of blind intubation is high, and above all, blind intubation may cause tissue damage. The transnasal and transoral fiberoptic intubation were then attempted, but both failed due to limited supraglottic space and safe duration of apnea, even under well chin lift and jaw thrust. An FOB provides a more flexible angle of view and guides the endotracheal tube. However, the limited space of oropharynx makes it difficult to insert the FOB through. A video laryngoscope provides panoramic view and eliminates the obstruction of the tongue. There are reports of a combined technique using the Glidescope video laryngoscope and FOB with difficult airways.[1] Iravani reported a case of fiberoptic intubation in a child with severe mandibular hypoplasia in which they held the tongue by surgical gauze and an FOB was inserted successfully.[2] Kim *et al*. used the I-gel™ supraglottic airway for ventilation and were able to achieve successful intubation with a cuffed tube by inserting FOB through the I-gel™ supraglottic airway.[3] This may be the safest and most effective method for difficult airway in pediatric patients with Goldenhar syndrome. Successful cases for these difficult intubations with other techniques were reported, such as pediatric GlideScope, Airtraq, wire-guided intubation, LMA,[4,5] and intubating LMA was also a good option. According to the anesthesia recommendations for patients suffering from Goldenhar syndrome, surgical airway equipment and experts should be ready.

Nevertheless, in our case, whether rapid sequence induction is the best choice remains a question. From a security perspective, maintaining spontaneous ventilation under the anesthesia of ketamine may have been a better option. Furthermore, the McGrath® video laryngoscope used was McGrath MAC 3, a McGrath Series 5, or Airtraq which may have provided better glottis exposure owing to the angle of its laryngoscope blade.

Anomalies due to Goldenhar syndrome include facial anomalies, ear abnormalities, ocular anomalies, vertebral anomalies, central nervous system malformations, cardiac malformations, and genitourinary anomalies. General anesthesia is recommended for pediatric patients with Goldenhar syndrome, as it is more independent of pediatric patients' cooperation and avoids urgent intraoperative intubation. Furthermore, intraspinal anesthesia may be difficult because of spine malformations. A comprehensive preoperative assessment is crucial for the preparation and management of anesthesia. Mallampati score, thyromental distance, and neck mobility are common assessment methods. In addition, presence of loose or protruding teeth, masses such as cystic hygroma or cysts in the mouth, mandibular size, and temporomandibular joint movement should also be taken into account. We also should obtain a clear medical history of any prior craniofacial surgery, previous anesthesia, or respiratory problems (snoring, sleep apnea, respiratory tract infections, etc.). A preoperative three-dimensional computed tomography provided a complete evaluation of the malformations. Moreover, the presence of the congenital heart defects, lung hypoplasia, vertebral anomalies, and genitourinary malformation should also be evaluated.

In summary, for pediatric patients with Goldenhar syndrome, a comprehensive preoperative assessment, adequate preparation, and alternative plans are important factors for successful airway management.

Declaration of patient consent

The authors certify that they have obtained the patient consent form. In the form, the patient's parents have given consent for images and other clinical information to be reported in the journal.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Fitzmaurice BC, Lambert BG. Failed fiberoptic intubation in a child with epidermolysis bullosa, rescued with combined use of the Glidescope®. Paediatr Anaesth 2016;26:455-6. doi: 10.1111/pan.12852.
2. Iravani M, Wald SH. Dexmedetomidine and ketamine for fiberoptic intubation in a child with severe mandibular hypoplasia. J Clin Anesth 2008;20:455-7. doi: 10.1016/j.jclinane.2008.03.012.
3. Kim YL, Seo DM, Shim KS, Kim EJ, Lee JH, Lee SG, *et al.* Successful tracheal intubation using fiberoptic bronchoscope via an I-gel™ supraglottic airway in a pediatric patient with Goldenhar syndrome – A case report. Korean J Anesthesiol 2013;65:61-5. doi: 10.4097/kjae.2013.65.1.61.
4. Heier JM, Schroeder KM, Galgon RE, Arndt GA. Wire-guided (Seldinger technique) intubation through a face mask in urgent, difficult and grossly distorted airways. Saudi J Anaesth 2012;6:292-4. doi: 10.4103/1658-354X.101226.
5. Khalil S, Vinh B. Successful intubation of a child with Goldenhar syndrome, who previously failed intubation, using an Airtraq. Paediatr Anaesth 2010;20:204-5. doi: 10.1111/j.1460-9592.2009.03223.x.