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Original Article

Airway management under general anesthesia for infants and young children with the first and second branchial syndrome featuring unilateral mandibular dysplasia: A case series of 8 children

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ABSTRACT

Objective: To investigate and summarize the airway management methods for infants and young children of the first and second branchial syndrome featuring mandibular dysplasia, and to evaluate the auxiliary effect of direct laryngoscope and video laryngoscope during tracheal intubation.
Methods: From March 2017 to March 2022, 8 cases with the first and second branchial syndrome featuring absent or hypoplastic mandibular ascending ramus that underwent cleft palate repair or transverse facial cleft repair under general anesthesia were retrospectively reviewed and summarized. The information such as demographic data, preoperative airway assessment, mask ventilation effect, anesthesia method, anesthesiologist's evaluation of laryngoscope exposure and intubation, operation method, operation time, and extubation time was collected.
Results: The median age of the 8 children was 12 months; none of them had limitation of mouth opening, 4 had snoring during sleep, 2 had unilateral absence of the ascending ramus of the mandible, and 6 had partial absence. Of the 8 children, 3 underwent cleft palate repair, and 5 underwent transverse facial cleft repair. During anesthesia induction, 1 case of mask ventilation was graded as Grade 2, and the other 7 cases were graded as Grade 1; the Cormack-Lehane (C-L) grade of glottic exposure by direct laryngoscope was graded as Grade 3 (3 cases) and Grade 4 (5 cases), and the C-L grade by video laryngoscope was graded as Grade 1 (4 cases) and Grade 2 (4 cases). All the children completed video laryngoscope-assisted intubation successfully in one time. The extubation was completed smoothly, without complications related to anesthesia. The median operation time was 50 minutes, and the median time from end of operation to extubation was 240 seconds.
Conclusion: For anesthesia of infants and children with the first and second branchial syndrome, especially those with hypoplasia of the mandible, a comprehensive preoperative assessment is needed, and direct laryngoscope may lead to difficulty in glottic exposure, and adequate planning for difficult airway management is necessary.

1. Introduction

The first and second branchial syndrome is composed of a series of congenital malformations of facial structures derived from the first and second embryonic branchial arches, and the first pharyngeal pouch and the first branchial cleft intervening the arches, and the temporal bone primordium. The disease manifests as combined tissue deficiencies and hypoplasias of the face, external ear, middle ear and maxillary and mandibular arches. The most common features are transverse facial cleft, cleft lip with or without cleft palate, atresia of the pinna (the most severe cases can be absence of the internal, middle, and external auditory canals), and micrognathia [1]. The spectrum of disorders of facial structural dysplasia derived from the first and second branchial arches is so broad that clinicians sometimes use other diagnostic terms such as hemifacial microsomia (HFM) or craniofacial microsomia (CFM). If combined with other embryological deformities, such as ocular dermoid cysts, reauricular skin tags, spinal deformities, it is also called Goldenhar syndrome or oculo-auriculo-vertebral spectrum (OAVS) [2,3]. The incidence rate reported in the literature is about 1/5600–1/26,370, which is second only to cleft lip and palate in maxillofacial developmental malformations. The incidence in men is higher than that in women [2,4]. In addition to maxillofacial malformations, children may also have congenital heart disease, genitourinary malformations (e.g.renal

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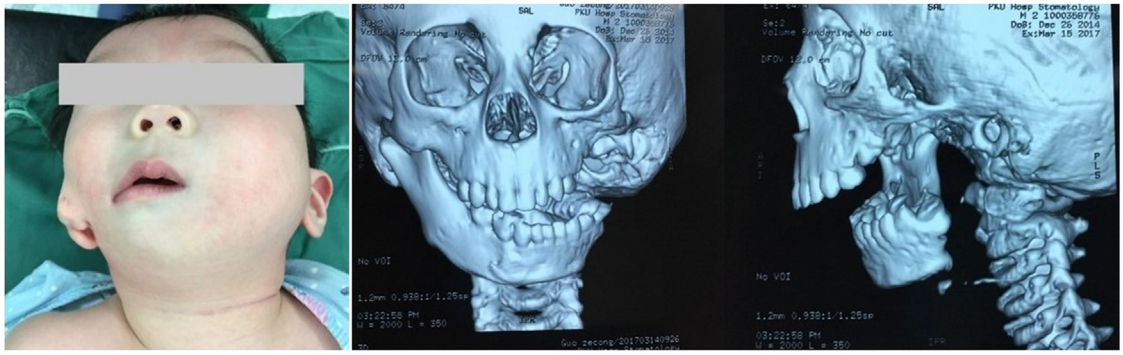

Fig. 1. Representative case, a 24-month male child. (a) Facial photo show right Transverse facial cleft, microtia and micrognathia; (b) 3D CT reconstruction show complete absence of left mandibular ramus.

hypoplasia, and ureteral malformations), and central nervous system abnormalities (for example, ventriculocele, and hydrocephalus).

Severe deformities of the maxillofacial region of the first and second branchial syndrome may affect children's breathing, facial muscle movement, appearance, language, and eating. Due to the possibility of abnormal development of other organs, multidisciplinary teams composed of pediatrics, maxillofacial surgery, and cardiac surgery should provide evaluation and individualized sequential treatment for children, some of whom may need surgery from infancy [5].

Because patients with the first and second branchial syndrome may have mandibular hypoplasia or even abnormal temporomandibular joints and glossoptosis, some children have obstructive sleep apnea [6] and anesthesiologists may encounter difficulties during tracheal intubation. However, there are no studies of airway management in such patients currently. In this article, the airway management of 8 infants and young children with first and second branchial syndrome featuring unilateral absent or hypoplastic mandibular ascending ramus was summarized, and the auxiliary effect of direct laryngoscope and video laryngoscope during tracheal intubation of these patients was evaluated.

2. Materials and methods

From March 2017 to March 2022, 8 infants and young children diagnosed with first and second branchial syndrome featuring absent or hypoplastic mandibular ascending ramus (Figs. 1 and 2) underwent cleft palate repair or transverse facial cleft repair in Peking University Hospital of Stomatology. The information such as demographic data, preoperative airway assessment, mask ventilation effect, anesthesia method, anesthesiologist's evaluation of laryngoscope exposure and intubation, operation method, operation time, and extubation time was collected.

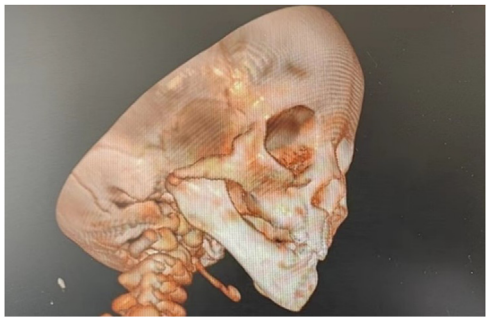

Fig. 2. 3D CT reconstruction of another child show partial absence of right mandibular.

Anesthesia induction was conducted by inhalation of 6–8 L/min O_2 and 4–8% sevoflurane. After unconsciousness, intravenous access was established, and 1–2 mg/kg propofol was administered depending on the depth of anesthesia and hemodynamics. After spontaneous respiration disappeared, the glottis was exposed by using a common laryngoscope. The anesthesiologists chose direct intubation or tracheal intubation by replacing with a video laryngoscope (UE, VL300M) depending on children's conditions and their own experience. During the operation, anesthesia was maintained by sevoflurane (2–4%) and Air/O_2=50% under spontaneous respiration. The anesthetic concentration was adjusted according to the heart rate, blood pressure, tidal volume (>4 ml/Kg) and end-tidal partial pressure of carbon dioxide ($EtCO_2$). The anesthesia monitoring included electrocardiogram, SPO_2, axillary temperature, $EtCO_2$, and meanwhile, the tidal volume, respiratory rate, respiratory amplitude, and lip color were observed. At the end of the operation, when the children were awake, the tracheal cannula was removed after sufficient sputum suction, and children were transferred to the anesthesia recovery room.

The mask ventilation classification (Han R et al.) was as follows, Grade 1: ventilated by mask; Grade 2: ventilated by mask with oropharyngeal airway/other adjuvant, with or without muscle relaxants; Grade 3: difficult mask ventilation (inadequate, unstable or requiring two practitioners), with or without muscle relaxants; Grade 4: unable to mask ventilation, with or without muscle relaxants [7]. The Cormack-Lehane (CL) classification was used to describe glottic exposure by laryngoscope, Grade 1: full view of the glottis; Grade 2: partial view of the glottis or arytenoids; Grade 3: only epiglottis visible; Grade 4: neither glottis nor epiglottis visible.

3. Results

The median age of the 8 children was 10.5 months; 6 children were male, and none of them had limitation of mouth opening or limitation of neck extension, 4 had snoring during sleep, 2 had unilateral absence of the ascending ramus of the mandible, and 6 had partial absence. The basic information of children was shown in Table 1. Three children underwent cleft palate repair, and 5 children underwent transverse facial cleft repair. During anesthesia induction, 1 case of mask ventilation was graded as Grade 2, ventilated by mask with oropharyngeal airway, and the other cases were graded as Grade 1; the Cormack-Lehane (C-L) grade of glottic exposure by direct laryngoscope was graded as Grade 3 (3 cases) and Grade 4 (5 cases), and the C-L grade by video laryngoscope was graded as Grade 1 (4 cases) and Grade 2 (4 cases) (Fig. 3). All the children completed video laryngoscope-assisted intubation successfully in one time. The extubation was completed smoothly, without complications related to anesthesia. The median operation time was 50 minutes, and the

Table 1
Basic information of the cases.

Patient	Age (m)	Sex	Height (cm)	Weight (kg)	Snore	Micrognathia	Mandibular ramus growth	Ear deformity	Eye anomaly	Heart deformity
1	12	M	70	7.5	Y	Y	Right, complete absence	Microtia	N	PDA
2	24	M	90	12.5	Y	Y	Left, complete absence	Microtia, bilateral accessory ear	L, dermoid cyst	ASD
3	3	M	55	7	N	Y	Right, Partial absence	Accessory ear	N	N
4	22	M	80	10	Y	Y	Left, Partial absence	Accessory ear	N	N
5	10	F	71	8	N	Y	Left, Partial absence	Microtia	N	N
6	4	F	67	7	N	Y	Left, Partial absence	Accessory ear	N	VSD
7	10	M	65	9	Y	Y	Right, Partial absence	N	N	N
8	11	M	70	8.5	N	Y	Left, Partial absence	N	N	N

M: Male; F: Female; Y: Yes; N: No; PDA: Patent Ductus Arteriosus; ASD: Atrial Septal Defect; VSD: Ventricular Septal Defect.

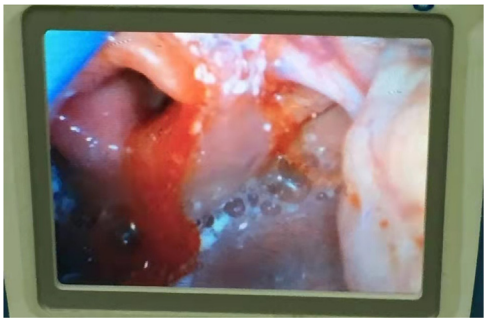

Fig. 3. C-L Grade 2 under video laryngoscope.

median time from end of operation to extubation was 240 seconds (Tables 2 and 3).

4. Discussion

The first and second branchial syndrome is the maxillofacial deformity second only to cleft lip and palate in the incidence rate. Changes in gene and protein expression and activation during facial development determine the manifestation and severity of maxillofacial defects. Mild patients with mandibular dysplasia have the manifestations of mildly flattened condylar head, and sever patients have the manifestations of condylar process, ascending ramus of the mandible, and even temporomandibular joint fossa hypoplasia [8]. In this study, preoperative three-dimensional reconstruction of the maxillofacial region of the included 8 children showed that 2 cases had ascending mandibular ramus, condylar process and complete absence of temporomandibular joint, 6 cases had incomplete development of the ascending ramus of the mandible and no connection with the temporal bone. According to the Kaban-Pruzansky diagnostic classification [9], they respectively belonged to Pruzansky type IIB (hypoplasia of the mandibular ramus, with obvious abnormal shape and location, located medially and anteriorly, without connection with the temporal bone) and type III (ascending mandibular ramus, condylar process and absence of temporomandibular joint). Jaw and neuromuscular dysplasia may lead to upper airway obstruction in children. In a retrospective study of 755 patients with CFM, 17.6% of them were diagnosed with obstructive sleep apnea (OSA), which is higher than the prevalence rate of OSA in healthy populations described in the literature [6], and the number of patients of Pruzansky type IIB/III or bilateral involvement who were diagnosed with OSA was significantly higher than that of patients of Pruzansky type I/IIA. In this study, all of the 8 patients had the manifestation of micromandible before surgery, and 4 of them had snoring during sleep as stated by their family members. Although no sleep monitoring was performed, the infants had no signs of asphyxia or hypoxia after birth, and did not receive OSA-related treatment, such as continuous positive airway pressure (CPAP) or placement of a nasopharyngeal airway. The cervical spine is also one of the concerns of

Table 2
Airway management of the cases.

Patient	Mask ventilation classification	Oropharyngeal airway	Cormack-Lehane (CL) classification		Trachea cannula procedure		
			Direct laryngoscope	Video laryngoscope	Technique	Tube type	Times
1	2	Y	4	2	Video laryngoscope	4# with capsule	1
2	1	N	3	1	Video laryngoscope	4# with capsule	1
3	1	N	4	1	Video laryngoscope	4# without capsule	1
4	1	N	4	2	Video laryngoscope	4# with capsule	1
5	1	N	4	2	Video laryngoscope	4# with capsule	1
6	1	N	3	1	Video laryngoscope	4# without capsule	1
7	1	N	4	2	Video laryngoscope	4# with capsule	1
8	1	N	3	1	Video laryngoscope	4# without capsule	1

Table 3
Outcomes of the operation and anesthesia of the cases.

Patient	Surgery	Operation time(min)	Postoperative extubation time(s)	Complication
1	Cleft palate repair	50	300	N
2	Transverse facial cleft repair	55	200	N
3	Transverse facial cleft repair	105	240	N
4	Transverse facial cleft repair	40	210	N
5	Cleft palate repair	30	300	N
6	Transverse facial cleft repair	60	300	N
7	Cleft palate repair	40	180	N
8	Transverse facial cleft repair	50	240	N

anesthesia intubation in such children. Ingeborg Barisic et al. [2] found that about 24.3% of children with OAVS had spinal deformities, which might occur in all spinal segments, but were more common in the cervical spine. Only a few for diffuse upper cervical fusion [10], children may have limited cervical rotational movement. However, in this study, the preoperative maxillofacial CT 3D reconstruction of the children showed no obvious abnormal cervical deformity, and the anesthesiologist did not find obvious limitation of neck movement of the children.

Based on the above clinical characteristics of the maxillofacial region of patients with the first and second branchial syndrome, it should be alert that children may have difficulty in ventilation or intubation during general anesthesia, and a plan for difficult airway management should be prepared. The principle for ensuring the safe anesthesia induction in difficult airways is to maintain spontaneous respiration, which can be achieved by intubation techniques for adult patients staying awake. For pediatric patients, M Cardwell and RWM Walker [11] recommended inhalation induction with 100% oxygen, sevoflurane or halothane as the first option, and muscle relaxants must be avoided before ensuring safe airway, to avoid "no ventilation, no intubation". In this study, the anesthesiologists all chose pure oxygen and sevoflurane inhalation induction for 8 children. In order to ensure the depth of anesthesia during the intubation process, 1 −2 mg/kg of propofol was given after open intravenous infusion. The study by Jin et al. [12] showed that, 93.8% of patients had Grade I mask ventilation, 5.7% had Grade II ventilation (with oropharyngeal airway), and 0.3% has grade III ventilation (requiring 2 physicians). In this study, 1 patient (12.5%) had difficulty in mask ventilation during the induction process, which improved significantly after placement of oropharyngeal airway. For young patients with the first and second branchial syndrome featuring severe mandibular involvement, the children's respiratory indexes such as tidal volume, respiratory rate, respiratory amplitude, SPO_2, $EtCO_2$, lip color should be carefully observed during anesthesia induction, and oropharyngeal airway and even laryngeal mask should be prepared [13].

In the study by Jin et al. [12], the proportion of C-L grade $\geq$ 3 of glottic exposure by direct laryngoscope was 38.9%. In this study, anesthesiologists exposed the glottis by direct laryngoscope. Results showed C-L grade $\geq$3 in 8 children (100%), of which 3 cases belonged to C-L grade 3 and 5 cases belonged to C-L grade 4, suggesting that glottic exposure by direct laryngoscope may be more difficult for infants and young children with the first and second branchial syndrome featuring unilateral absent or hypoplastic mandibular ascending ramus. In the process of exposing the glottis by video laryngoscope, it is not needed to lift the epiglottis excessively to overlap the three axes of the mouth, pharynx and larynx. Therefore, the irritation and damage to the larynx are reduced while a wide field of view is available, improving the success rate and safety of tracheal intubation. In this study, the anesthesiologist chose to use a video laryngoscope (UE, VL300M type) when the glottic exposure by direct laryngoscope was difficult. The C-L grading under the video laryngoscope improved, with 2 cases of Grade 1, 3 cases of Grade 2, and one-time intubation was completed successfully, which was basically consistent with the conclusions made by Jin Xu, et al [12]. In addition, intubation by video laryngoscope has less impact on the patient's hemodynamics [14,15]. Therefore, video laryngoscope can be considered as the first choice for airway management of children with first and second branchial syndrome, especially those with markedly abnormal mandibular development. Although all the cases were successfully intubated with a video laryngoscope in this study, there was a case report that one child with Goldenhar syndrome underwent surgery at 5 months of age, and retrograde endotracheal intubation was used after failure of 5 times of conventional intubation [16]. Shukry M [13] reported an 8-year-old child with Goldenhar syndrome who had difficulty in ventilation after induction of general anesthesia. After a laryngeal mask was placed to ensure ventilation, direct laryngoscopic or fiberoptic bronchoscopic (FFB) intubation failed, and finally the intubation was performed successfully by Shikani. For such patients, sufficient airway tools, including laryngeal mask, fiberoptic bronchoscope, and Shikani, should be prepared before anesthesia.

Patients, especially infants and young children with the first and second branchial syndrome, may have developmental retardation, cardiac malformation and difficult airway. In this study, 3 children were complicated with congenital heart disease. Intraoperative maintenance of heart rate is the key to maintaining cardiac output, and it is required to maintain spontaneous respiration. In order to reduce the risk of postoperative apnea, intravenous infusion of medicine (muscle relaxants, opioids) is avoided during the intraoperative anesthesia maintenance phase. As a commonly used inhalation anesthetic, sevoflurane has the characteristics of rapid induction, maintaining of spontaneous respiration during operation, stable anesthesia, and quick recovery, which is suitable for anesthesia in such children [17,18]. However, in order to prevent hypercapnia induced by CO_2 accumulation of spontaneous respiration, $EtCO_2$ monitoring was required during the operation and manual assisted respiration should be given when necessary. For manual assisted respiration, the dosage of anesthetics can be flexibly adjusted by observing the respiratory frequency and tidal volume, to effectively avoid the problems of insufficient tidal volume and prolonged wake-up time, thereby accelerating wake-up and recovery of respiratory functions [19,20].

In this study, extubation was performed successfully in 8 children during the anesthesia recovery period, and there were no complications such as laryngospasm and post-extubation respiratory depression. But attentions should be paid to the respiratory management of the children during the extubation stage. Studies have showed that, the abnormality of the throat in children with OAVS and the severity were not related to the severity of the mandibular deformity [21]. Nasopharyngoscopy showed abnormalities, including narrowing of the anteroposterior diameter of the trachea, asymmetric movements of the laryngeal or cricoid cartilage, and shortening or irregular shape of the epiglottis, and arytenoid cartilage covering excess mucosa. In this study, there was 1 child whose arytenoid cartilage was covered by a large amount of excess mucosa, protruded forward on expiration to completely cover the glottis, and abducted on inspiration. Although the child had no history of dyspnea, obvious difficult breathing occurred after anesthesia extubation. Therefore, for children with the first and second branchial syndrome, preoperative nasopharyngoscopy can be considered. In the extubation stage, the tracheal tube must be removed after ensuring that the child is awake and his/her breathing returns to normal and conform to the indications for extubation. At the same time, oral secretions and blood must be sucked to avoid aspiration or irritation of the airway, which may cause laryngospasm. And the corresponding nasopharyngeal airway, oropharyngeal airway, laryngeal mask and other ventilation equipment should be well equipped, to get ready for re-intubation.

5. Conclusion

In conclusion, infants and young children with the first and second branchial syndrome, especially those with mandibular hypoplasia, should receive a comprehensive preoperative evaluation. During the intubation under general anesthesia, glottic exposure by direct laryngoscope may be difficult, while the success rate by video laryngoscope is high. It is necessary to make a plan for difficult airway management and prepare for a variety of airway tools.

Statement

Written informed consent was obtained from the patient for the publication of any potentially identifiable images or data included in this article.

Declaration of Competing Interest

The authors declare no conflict of interest.

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