

Successful use of C-MAC™ Miller blade in a neonate with Goldenhar syndrome and cleft lip with cleft palate : a case report

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Abstract: A 20 days old neonate with Goldenhar Syndrome and cleft lip with cleft palate was scheduled for ocular surgery. We discuss the management of the anticipated difficult airway in the patient using a C-MAC™ (Karl Storz, Tuttingen, Germany) Miller blade. A thorough pre-anesthetic evaluation and meticulous planning for airway management is of utmost importance in neonates with this or comparable syndromes.

Keywords- Goldenhar syndrome; cleft lip; cleft palate; C-MAC

INTRODUCTION

Goldenhar syndrome is a rare syndrome, also known as “oculo-auriculo-vertebral” dysplasia. Cleft lip-palate has a frequent association with the syndrome along with other facial deformities leading to airway problems (1). A difficult airway along with systemic involvement poses a challenge for the anesthesiologist. In neonates with a difficult airway, the primary goal of anesthesia for urgent surgery is early airway protection without oropharyngeal and laryngeal trauma (2). We present the airway management of a neonate with facial deformity and cleft lip-palate undergoing urgent ocular surgery.

CASE REPORT

A 20 days old male neonate weighing 2.5 kg and suffering from the Goldenhar syndrome was scheduled for left eye lipodermoid excision, lid reconstruction and penetrating keratoplasty. The child was born full term by normal vaginal delivery and perinatal history was uneventful. On facial examination, the child had a left eyelid coloboma, limbal dermoid, keratopathy and left pre-auricular tags. His airway examination revealed grade IV cleft lip-palate and a receding mandible with normal neck movements. On auscultation, a systolic murmur was present with bilateral vesicular breath sounds. Routine blood investigations were normal. X-ray chest and spine and ultrasound abdomen-pelvis were normal. Echocardiography revealed a 2



mm patent foramen ovale with left to right shunting and normal bi-ventricular function.

Anesthetic management

Parental consent was obtained for surgery and to publish details including a picture of the patient.

As the neonate was exclusively breastfed, 4 hours fasting was advised and ensured before surgery. No pre-medication was administered. Our difficult airway cart was prepared. After arrival of the patient in the operation theatre, a five lead electrocardiogram (ECG), non-invasive blood pressure (NIBP) and pulse oximetry monitors were attached. Pre-induction, a 24G intravenous cannula

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was inserted in the left foot. Induction of anesthesia was performed with increasing sevoflurane concentrations (2-8%) in 4 l/min 100% oxygen using a pediatric breathing circuit. Assisted mask ventilation was difficult due to the receding mandible and was possible only with jaw thrust and a two hand technique. After achieving an adequate depth of anesthesia, an exploratory videolaryngoscopy was performed using a C-MAC™ size 0 Miller blade, keeping the blade towards the right angle of the mouth. Intra-oral anatomy was distorted, however, epiglottis and arytenoids were visible. Assisted ventilation was resumed to increase depth of anesthesia and videolaryngoscopy was performed again using the same blade and technique. Now, a Cormack-Lehane grade 2b could be obtained, which was improved to 2a by optimal external laryngeal manipulation. The trachea was intubated with a styletted uncuffed endotracheal tube (ETT, inner diameter 3.0mm). After confirmation of ETT placement, atracurium 1.25 mg and fentanyl 5 µg were administered intravenously. Anesthesia was maintained with sevoflurane in oxygen and air (FiO₂ 0.5). Intermittent boluses of fentanyl 1.25 µg and atracurium 0.5 mg were repeated as required. Paracetamol 7.5 mg/kg was administered intravenously. Surgery lasted for 2 hours and the intraoperative period was uneventful. At the end of surgery, residual neuromuscular blockade was reversed with neostigmine and glycopyrrolate. The trachea was extubated when the child was fully awake. The postoperative period was uneventful. The child was transferred to the ward after 4 hours and discharged on the second postoperative day.

DISCUSSION

The Goldenhar syndrome was first described by Dr. Maurice Goldenhar in 1952 and classified as “oculo-auriculo-vertebral” dysplasia by Gorlin *et al.* (3, 4). Our patient had various components of the Goldenhar syndrome as described by Feingold and Baum in their diagnostic criteria (5).

The anesthetic management was challenging due to an anticipated difficult airway, including retrognathia with grade IV cleft lip and palate which can lead to difficult mask ventilation and intubation. Since assisted mask ventilation was difficult in our case only possible using the two hand technique and jaw thrust, we planned to maintain spontaneous ventilation until the airway was secured. Tremlett *et al.* also suggested that in children with facial anomalies including cleft lip-palate, maintaining spontaneous ventilation is the safest approach (6).

We used the C-MAC™ for an exploratory laryngoscopy since videolaryngoscope (VLS) blades require less lifting force. Nienaber *et al.* also suggested that the use of VLS in pediatric patients with split palate and lip defects has the additional advantage of not dislodging into the cleft, as the VLS handle only needs to be lifted and not tilted as anteriorly as in direct laryngoscopy (7).

The tendency of the laryngoscope blade to fall into a wide cleft may be overcome in two ways: packing the cleft with a small gauze roll or inserting the laryngoscope from the side of the mouth rather than from midline (6). Hence we chose to insert the VLS Miller blade from the right corner of the mouth. Since we could visualise the arytenoids during the exploratory laryngoscopy, we planned to proceed with the same technique using the C-MAC™ pediatric size 0 Miller blade. We were successful in tracheal intubation without experiencing difficulties after optimal external laryngeal manipulation. In a prospective study, Mutlak *et al.* also concluded that videolaryngoscopy using the C-MAC-Miller blade and the Glidescope-GVL 2 in infants with cleft lip-palate significantly improved viewing of the glottis and reduced the rate of difficult laryngoscopy (8).

Our primary plan was to use VLS due to previous reports of failure of direct laryngoscopy (DL) in children with cleft lip-palate necessitating the use of other devices. Milne *et al.* had to use the Glidescope® (Bothell, Washington, USA) after unsuccessful intubation with DL in a nine year old girl with the Goldenhar syndrome (9). A recent Cochrane review conducted in adults concluded that videolaryngoscopes may improve the success of tracheal intubation, particularly when the patient has a difficult airway (10).

We preferred the use of the C-MAC™ over fiberoptic intubation as the latter technique takes longer time to secure the airway and neonates have a low threshold for desaturation.

We did not select supraglottic devices (SGD) as a definitive airway device in view of neonatal age, long duration of surgery and the risk of device displacement under the drapes in ocular surgery. However, we were prepared to use SGD as a conduit for intubation and rescue device for ventilation.

In conclusion, patients with Goldenhar syndrome must undergo a thorough pre-anesthetic evaluation to assess systemic and airway involvement. Difficult airway management in neonates requires identification of the problems and meticulous planning. Choosing an appropriate method and airway device is a key factor to avoid major airway-related complications in such patients.

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