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Goldenhar Syndrome: A Perpetual Airway Challenge for the Anaesthesiologist

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Abstract

Patients with Goldenhar syndrome present as difficult airway to the anaesthesiologist due to various craniofacial anomalies like hemifacial microsomia, retrognathia and high arched palate. Also, these patients often undergo repeated surgeries for their various deformities. A, 4 year old female with Goldenhar syndrome and diagnosed as Right microphthalmos was posted for second stage lid reconstruction surgery. Previous anaesthesia records revealed no history of difficult intubation. During second stage, difficult intubation was encountered and eventually patient's trachea was successfully intubated with C-MAC® D-blade videolaryngoscope.

Keywords: Goldenhar; D blade CmacÂ®; Difficult airway

Introduction

Difficult mask ventilation as well as difficult intubation has been reported in patients with Goldenhar syndrome attributed to craniovertebral anomalies, micrognathia, retrognathia and high arched palate which tend to progress with age [1,2]. Various techniques have been used in securing a definitive airway in this paediatric population ranging from tracheostomy in earlier era to fibreoptic bronchoscopy, till date. We present a case of successful airway management with C-MAC® D-blade videolaryngoscope in a child with Goldenhar syndrome.

Case report

A, 4 year old girl (15 kg ASA I), with Goldenhar syndrome presented for pre -anaesthetic evaluation for second stage lid reconstruction surgery with mucous membrane grafting and right epilimbal dermoid excision. She had been earlier operated at three years of age for the same condition. Her previous anaesthesia records showed difficult mask ventilation with Cormack-Lehane Grade II on intubation with McCoy blade #1. On history taking, she was younger of the three siblings with no family history of such disorders. She was a full term vaginal delivery at a peripheral health centre. Her developmental milestones were normal with no history suggestive of congenital heart disease. On airway examination, she had micrognathia, retrognathia, left mandibular hypoplasia with high arched palate. Dentition was irregular. She had right preauricular tag with nasal tag. Neck movements were normal. Mouth opening was greater than patient's two fingers. Rest of systemic examination was within normal limits. Preoperative haemoglobin was 10.5 gm/dl with normal platelet count. Preoperative echocardiography showed no cardiac abnormality. X-ray neck was normal with no craniovertebral anomaly.

Patient was kept 6 hours fasting with difficult airway management explained to parents and high risk consent taken. Operation theatre was prewarmed and difficult airway cart was kept ready with appropriate sized oral airway, proseal LMA, bougie, stylet and C-MAC® D-blade videolaryngoscope. A 22G catheter was secured intravenously in the preholding area.

After standard ASA monitors were attached, anaesthesia was induced with fentanyl 2µg/kg and propofol 30mg intravenously. Bag mask ventilation revealed a leak for which gauze on the side of the left cheek was used to secure adequate seal which was verified by chest rise and end–tidal CO_2 end-tidal CO_2. When adequate depth of anaesthesia was achieved, a LMA-ProSeal size #1.5 was inserted, but proper placement and adequate ventilation could not be achieved, despite readjustment of the device. Succinylcholine 1 $mg.kg^{-1}$ was administered subsequently. Direct laryngoscopy using a Macintosh blade size #1 blade revealed non visualization of epiglottis (Cormack-Lehane Grade IV). C-MAC® D-blade videolaryngoscope was used, and with external laryngeal manipulation posterior commissure was visualized.

The trachea was subsequently intubated with an uncuffed endotracheal tube size #5.0 and bilateral air entry was confirmed. Anaesthesia was maintained with vecuronium 1.5 mg, oxygen-nitrous oxdie and 1 vol% isoflurane. Intraoperative haemodynamic parameters (blood pressure 90/60 mm Hg; pulse rate of 90/min and SpO_2 99%) were all stable and within normal limits. Surgery was completed after 55 minutes and reversal of neuromuscular blockade (glycopyrrolate 0.15 mg and neostigmine 0.75 mg) resulted in spontaneous respiration, which allowed extubation. The postoperative period was uneventful.

Discussion

Carl Ferdinand Von Arlt in 1881 first described the anomalous development of first and second brachial arches resulting in hemifacial microsomia. In 1952, Goldenhar associated these hemifacial anomalies with or without epibulbar dermoids, auricular appendages and auricular fistulas [3]. This was further redefined as oculo-auriculo-vertebral spectrum by Gorlin in 1962 [4]. Feingold and Baum' listed criteria for diagnosis of this syndrome requires patient having eye anomalies like lipoma, lipo-dermoid, epibulbardermoid along with at least one of cardiac, ear and vertebral colon defects [5]. With an incidence between 1/3000 and 1/5000 live births, it has a male preponderance in the ratio of 3:2 [6]. Most cases are sporadic in occurrence with relatively few having familial inheritance.

Numerous cases of Goldenhar's syndrome present with difficult airway, as reported in the literature with incidence around 39.5% [7]. Difficult mask ventilation occurs due lateral or mid-cheek extension of soft tissue cleft in the affected side [8]. In our patient mask ventilation was difficult but could be achieved successfully with the help of gauze which provided an adequate seal.

Difficult intubation in Goldenhar syndrome arises from a combination of asymmetrical mandibular hypoplasia, hemifacial microsomia, tracheal deviation to one side, and craniovertebral abnormalities [1,2]. In our patient previous attempt at intubation had been successful with Mcintosh blade but this time it revealed a Cormack-Lehane Grade IV. It has been reported in literature that the incidence of difficult intubation increases as the child, with a Goldenhar's syndrome, grows, due to bilateral clefts and retrognathia [9].

These patients are of particular importance to the anaesthesiologist as they come for repeated surgeries for their various anomalies with an anticipated difficult airway. As most of these patients belong to paediatric age group they have a predisposition to hypoxia owing to decreased physiological reserves like functional residual capacity and total lung capacity.

In our patient, endotracheal intubation was successful with direct laryngoscopy at the initial surgery, but the second time it proved difficult with a Cormack-Lehane Grade IV.

The availability C-MAC® D-blade videolaryngoscope helped in improving the laryngoscopy view of the glottis, resulting in successful intubation.

C-MAC® D-blade videolaryngoscope use CMOS technology in the distal lens which guarantees a uniform illuminated view of oropharynx with at least 60 degree field of vision without fogging or requiring the traditional sweeping of tongue. The D-blade if of particular help in patients with a Cormack-Lehane III and IV which due to its elliptical tapering distally rising blade shape improves vision, providing a rapid and compact alternative in difficult airway situations with a relatively shorter learning curve.

Conclusion

In conclusion, we emphasize that, in patients with Goldenhar's syndrome, even with a previous history of successful intubation, the anaesthesiologist has to foresee the eventuality of difficult airway and be prepared for its management.

Written consent was obtained from the patient's mother and she was explained the contents of the case report in the language best understood by her.

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