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Case report

Edward's syndrome: A rare cause of difficult intubation-utility of left molar approach



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KEYWORDS

Edward's syndrome; Left molar approach; Difficult intubation **Abstract** Edward's syndrome (trisomy 18) is an autosomal abnormality with dysmorphic face, visceral deformities and delayed mental and motor development including congenital heart disease. Challenges may arise during mask ventilation, laryngoscopy and/or intubation of the trachea due to dysmorphic face. Difficult airway cart should be kept ready. Left molar approach using a standard Macintosh blade improves the laryngoscopic view in patients with difficult midline laryngoscopy. We hereby present a case report of a 2 year old male child with Edward's syndrome posted for evacuation and drainage of brain abscess, intubated successfully using left molar approach.

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1. Introduction

Congenital syndromes are uncommon cause of difficult airway. The airway management of a paediatric patient with a syndrome associated with difficult airway is an ongoing challenge for the anaesthesiologist. Effective airway management includes anticipating and planning for problems. Difficulties frequently occur as the result of patient characteristics that interfere with bag mask ventilation, laryngoscopy and/or intubation of the trachea. Edward's syndrome (trisomy 18) is a chromosomal anomaly in which the patients are dysmorphic with multiple organ defects including congenital heart disease and delayed motor and mental development [1]. We hereby present a case report of a 2 year old male child with Edward's syndrome posted for evacuation and drainage of brain abscess, intubated successfully with left molar approach.

2. Case report

A 2 year old male child weighing 7 kg was posted for evacuation and drainage of brain abscess. He was a vaginally delivered full term born child weighing 1.9 kg during birth. He presented with fever and vomiting of 3 day duration. On general physical examination, he had dysmorphic face, flat nose, receding chin, overlapping fingers of limbs and omphalocele (Figs. 1 and 2). Localised swelling was present over left parietooccipetal region of the skull. Mental development and developmental milestones were delayed. Pulse was 112/min. Respiratory and cardiovascular systems were unremarkable. This child was diagnosed as a case of Edward's syndrome (partial trisomy 18) by the paediatrician on the basis of clinical features, further confirmed by Karyogram which demonstrated translocation of chromosome 18. Relevant blood biochemistry was within normal limits. Chest X Ray, ECG and echocardiography were normal. CECT head revealed multiple hypodense lesions with surrounding oedema in left parietooccipetal region with partial effacement of right lateral ventricle and dilatation of frontal horn of left lateral ventricle. Midline shift of 1.3 cm was seen towards right

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Figure 1 Showing dysmorphic face and flat nose.

side. Difficult intubation was anticipated in view of dysmorphic face. Difficult airway cart was kept ready. In the operating room, iv fluid was started through 24 G cannula in situ. Standard monitors were attached. Atropine 0.14 mg was given intravenously. Anaesthesia was induced with sevoflurane in oxygen. After assessing adequacy of ventilation, succinylcholine 10 mg was given to facilitate endotracheal intubation. Laryngoscopy was done with Miller blade but only epiglottis could be visualised even with optimal external laryngeal manipulation. Then we proceeded with left molar approach for laryngoscopy with a curved Macintosh blade which allowed visualisation of the glottis and trachea could be intubated successfully with endotracheal tube of internal diameter 4.5 mm confirmed by bilateral air entry and capnography. Analgesia was achieved with fentanyl. Anaesthesia was maintained with isoflurane, 67% N₂O in O₂ and vecuronium. Surgery lasted for one hour. At the end of surgery, residual neuromuscular blockade was reversed with atropine and neostigmine and trachea was extubated with patient awake. Postoperative course was uneventful.

3. Discussion

Edward's syndrome is the second most common chromosomal anomaly next to down syndrome. The incidence of full trisomy 18 is 1 in every 3500–7000 birth and it leads to death at the newborn or infant stage in most cases. On the other hand, the incidence of partial trisomy 18 is very rare and is mild in comparison with full trisomy. Key points in the anaesthetic management of such patients are preoperative examination for congenital heart disease, difficulty in ventilation with mask and difficult endotracheal intubation [2].

We anticipated difficult airway in our patient because of dysmorphic face and kept difficult airway cart ready. Initially



Figure 2 Showing overlapping fingers.

laryngoscopy was done with conventional approach using Miller blade but only tip of epiglottis could be visualised even with optimal external laryngeal manipulation. Then we attempted laryngoscopy with Macintosh blade using left molar approach and after visualisation of glottis, trachea was intubated successfully.

Left molar approach is an unconventional technique, in which blade is inserted from the left corner of the mouth. This approach has been used in patients with glossopalatal ankylosis, massive neurofibroma of face and loose incisors [3–5]. Left molar approach using a standard Macintosh blade improves the laryngoscopic view in patients with difficult midline laryngoscopy. It has the advantage of utilising the maximum effect of molar approach because the larvngoscope blade can be brought fully to the left side of mouth with rest of the oral opening available for viewing the larynx [6]. This approach reduces the distance from the patient's teeth to the larynx and prevents maxillary structures coming into the line of view and hence it is advisable in difficult airway situations. The Macintosh blade is widely accepted because it enables quick, atraumatic larvngoscopy and lesser deviation of line of view from the ideal line than the Miller blade [7]. So we used Macintosh blade for left molar approach instead of Miller blade.

The unavailability of suitable size fiberoptic bronchoscope and the unreliability of blind nasal intubation especially in abnormal anatomical situations necessitate the anaesthesiologists to be familiar with alternative approaches. The left molar approach of laryngoscopy is quickest to be attempted without the need for extra equipment. The left molar approach could be part of the anaesthesiologists' armamentarium in cases of difficult laryngoscopy.

Conflict of interest

We have no conflict of interest to declare.

Source of support

Nil.

References

- [1] Courreges P, Nieuviarts R, Lecoutre D. Anaesthetic management of Edward's syndrome. Paediatr Anaesth 2003;13:267–9.
- [2] Ochiai N, Okutani P, Yoshimura Y. Anesthetic management of a patient with partial trisomy. J Anesth 1995;9:78–80, 18.
- [3] Batra YK, Rakesh SV, Menon P, Rao KLN. Successful airway management with left-molar laryngoscopy in glossopalatal ankylosis: a case report. Acta Anaesth Belg 2010;61:215–6.
- [4] Saini S, Bansal T. Anesthetic management of difficult airway in a patient with massive neurofibroma of face: utility of Rendell Baker Soucek mask and left molar approach for ventilation and intubation. J Anaesthesiol Clin Pharmacol 2013;29:271–2.
- [5] Sato N, Shingu K. Another reason to choose the left molar approach of laryngoscopy: to spare the incisor teeth. Anesthesiology 2002;96:1279.
- [6] Yamamoto K, Tsubokawa T, Ohmura S, Itoh H, Kobayashi T. Left-molar approach improves the laryngeal view in patients with difficult laryngoscopy. Anesthesiology 2000;92:70–4.
- [7] Marks RRD, Hancock R, Charters P. An analysis of laryngoscope blade shape and design: new criteria for laryngoscope evaluation. Can J Anaesth 1993;40:262–70.