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Case Report
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Anaesthetic Challenges in a Paediatric Patient with Escobar Syndrome-Difficult Airway and Postoperative Pneumothorax

Waleed Bin Ghaffar, Irfan Ul Haq, Arham Shahid, Samina Ismail
Department of Anaesthesiology, Aga Khan University Hospital, Karachi, Pakistan

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Abstract

Escobar syndrome (ES) is an autosomal recessive disorder characterised by the presence of pterygia in cervical, antecubital and popliteal regions. Anaesthesiologist encounter notable challenges in this syndrome, especially airway management due to associated malformations like cleft lip/palate, micrognathia, syngnathia, ankyloglossia, neck contracture, cervical spine fusion, limited neck extension and craniofacial dysmorphism. In addition to difficult airway, anaesthesiologist may encounter other perioperative challenges. Here, we report a paediatric patient with ES, who required general anaesthesia for laparoscopic inguinal hernia repair and orchidopexy. Initial attempt with video laryngoscope failed due to inability to visualise epiglottis. Subsequent attempt with fibreoptic bronchoscope also failed due to rapid decrease in oxygen saturation. He was finally intubated with fibreoptic bronchoscope along with oxygen insufflation with a 3 mm internal diameter polyvinylchloride endotracheal tube inserted nasally and connected to oxygen supply. Further perioperative challenges faced were intraoperative hyperthermia and postoperative pneumothorax with mediastinal shift. To the best of our knowledge, this is the first case reporting pneumothorax with mediastinal shift as a postoperative complication and use of oxygen insufflation through nasal tube during fibreoptic intubation in paediatric patient with ES.

Keywords: Multiple pterygium syndrome, Escobar syndrome, airway management, pneumothorax, mediastinal shift, anaesthesia management

Introduction

Escobar syndrome (ES) is a rare autosomal recessive disorder caused by mutation in CHRNG gene, on chromosome 2q.[1] Clinical hallmark of this disease is the presence of pterygia in cervical, antecubital and popliteal regions.[2] Anaesthesiologist encounter notable challenges in this syndrome, especially regarding airway management. These include micrognathia, syngnathia, ankyloglossia, neck contracture, cervical spine fusion, limited neck extension and craniofacial dysmorphism.[2] The disease is progressive, with the possibility of spine deformity along with restrictive lung disease.[3] Anaesthetic challenges and perioperative management of these patients have been mentioned in literature, but so far none has mentioned the occurrence of postoperative pneumothorax with mediastinal shift.

Case Description

A two and a half-year-old male child, weighing 9 kg, was planned for laparoscopic right inguinal hernia repair and bilateral orchidopexy. He was a diagnosed case of multiple pterygium syndrome, Escobar variant.

Preoperative assessment and preparation were done for the evaluation of associated congenital anomaly and systematic illnesses. Physical examination revealed frontal bossing, micrognathia, short webbed neck with limited extension, rocker bottom feet with prominent heels and firm contracture of bilateral elbows and knees. Patient had pectus excavatum and thoracic scoliosis with convexity toward right side. On the day of surgery, difficult airway trolley was prepared, and ENT team taken on board. Standard monitoring was used, including noninvasive blood pressure, electrocardiogram, end-tidal carbon dioxide, temperature and oxygen saturation (SpO_2). Inhalation induction was done with 8% sevoflurane in 100% oxygen. After assessing adequate bag mask ventilation, muscle relaxation was achieved with Cis-atracurium, and intubation was attempted with Storz C Mac video laryngoscope

Corresponding Author: Waleed Bin Ghaffar; E-mail: waleedghaffar@hotmail.com

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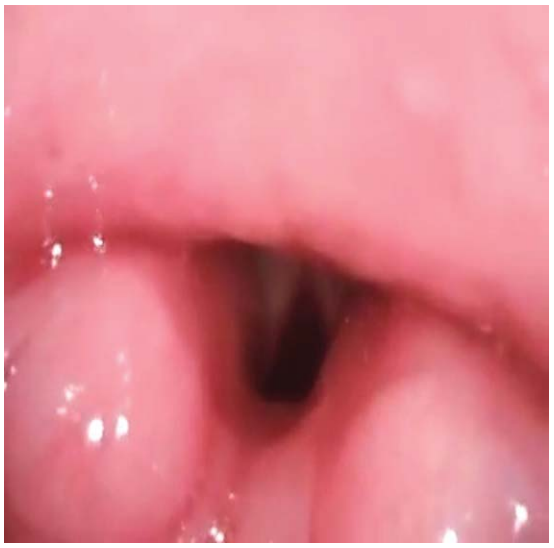

Figure 1. Fibreoptic view of vocals cords with oedematous arytenoids.

(Karl storz, Model 8403 ZX, Germany), which failed due to inability to visualise epiglottis. Subsequent attempt at intubation was done with paediatric fibreoptic bronchoscope (Karl storz endoscope, Germany), which failed due to rapid drop in SpO_2 associated with bradycardia. After managing with positive pressure ventilation and intravenous atropine, second attempt at fibreoptic intubation (FI) was planned with oxygen insufflation to prevent desaturation during the procedure. A 3 mm polyvinylchloride endotracheal tube (ET) was measured from tragus to nostril and inserted nasally and connected to circuit to provide oxygen at 5 L min^{-1}. Intubation was successful with a size of 4.0 mm oral ET. The larynx was found anteriorly, arytenoids were oedematous and only anterior commissure was visualised, as shown in Figure 1. After maintenance of airway, chest was auscultated to confirm bilateral air entry, and all pressure points were secured as shown in Figure 2. Temperature probe inserted nasally showed a temperature of 38°C. Inhalation anaesthesia was

Main Points

- Anaesthesiologist can encounter difficult airway as well as other unanticipated challenges while managing patients with Escobar syndrome.
- Kyphoscoliosis-induced restrictive lung disease can lead to functional airway abnormality with decrease oxygen reserve.
- Multiple management plan is needed to secure the airway in these patients.
- Availability of expertise and difficult intubation equipment including fibreoptic bronchoscope are required for anaesthesia provision.
- The presence of pulmonary hypoplasia, thoracic scoliosis and pectus excavatum can lead to spontaneous pneumothorax in ES.
- To avoid pneumothorax, it is important to ensure lung protective ventilation maintaining peak airway pressure below 25cm H_2O.

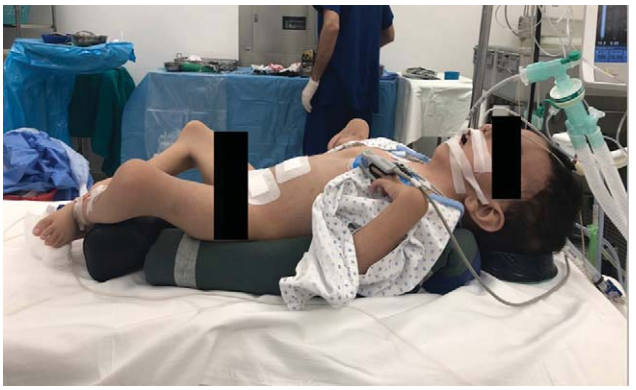

Figure 2. Patient positioning with secured pressure points.

turned off, circuit was changed and operating room temperature was lowered. As end-tidal carbon dioxide values were within the normal range (38-45 mmHg), surgery was allowed to proceed, and laparoscopy was performed to localise testis followed by inguinal hernia repair. Anaesthesia was maintained with intravenous propofol, nalbuphine and cisatracurium boluses guided by neuromuscular monitoring. Tidal volume of 6 mL kg^{-1} was achieved with a pressure of 18-22 cmH_2O. Intraoperative SpO_2 was between 92 and 96% with an inspired oxygen of 55-65%. On chest auscultation, there was bilateral equal breathing sound in all zones of lung. On completion of surgery, it was decided to transport the patient to paediatric intensive care unit (ICU), and extubation was deferred due to persistent hyperthermia and high inspired oxygen demand.

In ICU, increasing airway pressure with decreasing breath sound was noted on right side. Chest X-ray was done, which revealed a right-sided pneumothorax with mediastinal shift as shown in Figure 3. Immediately, a chest tube was inserted on the right side. The remaining ICU course was uneventful, and the patient was extubated within 12 hours, shifted out of ICU the following day and discharged on fourth postoperative day. Written informed consent was taken from the patient's parents before discharge from the hospital.

Discussion

This case report demonstrates that during the anaesthesia management of patients with ES, anaesthesiologist can encounter unanticipated challenges. The foremost challenge is securing the airway, and various ways of securing the airway have been mentioned in the literature, including laryngeal mask airway (LMA)-assisted fibreoptic-guided intubation,[4,5] optical stylet[6] and video laryngoscope.[7]

Video laryngoscopy is shown to provide high-resolution view of the paediatric difficult airway.[4,8] Therefore, our initial

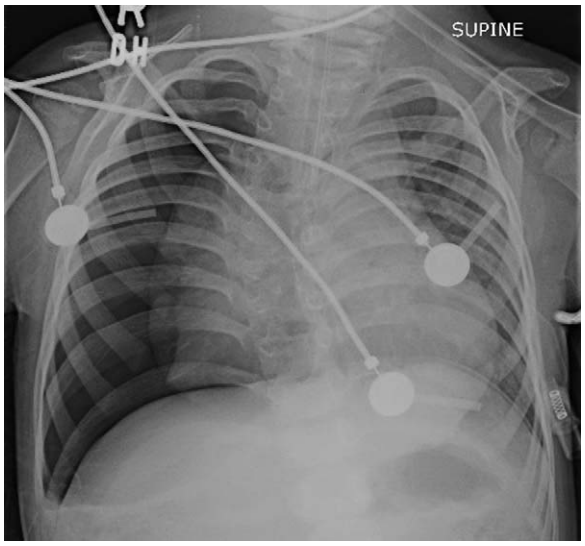

Figure 3. Right-sided tension pneumothorax with mediastinal shift.

plan was intubation with video laryngoscope; however, we were unsuccessful due to inability to visualise the epiglottis. Our subsequent plan was FI as previous reports have also advocated securing the airway in ES patients with FI.[5,9] However, our initial attempt at FI failed due to rapid drop in SpO_2, which could be due to anatomical and functional abnormality of alveoli secondary to thoracic scoliosis and pectus excavatum.[2,10] Previous case reports have also mentioned initial failure with FI and using Proseal LMA as conduit for fibreoptic bronchoscope.[4,5] We used a 3.0 mm nasal ETT as a nasal airway and connected it to oxygen port for oxygen insufflation, which helped in maintaining SpO_2, while trachea was successfully intubated by fibreoptic bronchoscope using oral route.

An association of ES with malignant hyperthermia has been mentioned in the literature.[11] However, it has been refuted by subsequent case reports, and the use of volatile agent is considered safe in these patients.[4,8,12] We encountered intraoperative hyperthermia without other signs of malignant hyperthermia, which settled postoperatively in ICU. Patients with ES have mutation in gamma subunit of AChR, that is why some authors have avoided muscle relaxants.[1] However, we used cis-atracurium and did not encounter extended duration of action.

Postoperative chest X-ray in ICU revealed pneumothorax with mediastinal shift, but our patient did not have haemodynamic instability. However, in ICU, he had decrease air entry on right side of the chest and increase airway pressure, which was not present in the operating room during surgery. The presence of mediastinal shift on X-ray does not necessarily imply that the patient has a tension pneumothorax as literature has shown radiological evidence of mediastinal shift without clinical features of tension pneumothorax.[13] The most possible cause of pneumothorax in this case seems to be endobronchial intubation, leading to one lung ventilation during transportation to the ICU. Other differential diagnosis of pneumothorax includes inadvertent increase in airway pressure during manual ventilation and introduction of laparoscopy ports. In addition, ES patients have pulmonary hypoplasia, thoracic scoliosis and pectus excavatum, which can be a cause of spontaneous pneumothorax.[14]

Conclusion

Managing patients with ES requires multiple management plan for securing the airway, ideally in a healthcare centre where resources are available in terms of expertise and equipment. These patients require vigilant perioperative monitoring for early diagnosis and management of any arising complication.

Informed Consent: Written informed consent was obtained from the guardian (parents) of patient who participated in this case.

Peer-review: Externally peer-reviewed.

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