Case Report

# Airway management in Escobar syndrome: A formidable challenge

**Address for correspondence:**
Dr. Souvik Chaudhuri,
Department of Anaesthesia,
Kasturba Medical College,
Manipal - 576 104,
Karnataka, India.
E-mail: souvik_mahe@yahoo.co.in

**Access this article online**

Website: www.ijaweb.org

DOI: 10.4103/0019-5049.123336

Quick response code

*Shaji Mathew, Souvik Chaudhuri, HD Arun Kumar, Tim Thomas Joseph*
Department of Anaesthesia, Kasturba Medical College, Manipal, Karnataka, India

## ABSTRACT

Escobar syndrome is a rare autosomal recessive disorder characterized by flexion joint and digit contractures, skin webbing, cleft palate, deformity of spine and cervical spine fusion. Associated difficult airway is mainly due to micrognathia, retrognathia, webbing of neck and limitation of the mouth opening and neck extension. We report a case of a 1 year old child with Escobar syndrome posted for bilateral hamstrings to quadriceps transfer. The child had adequate mouth opening with no evidence of cervical spine fusion, yet we faced difficulty in intubation which was ultimately overcome by securing a proseal laryngeal mask airway (PLMA) and then by intubating with an endotracheal tube railroaded over a paediatric fibreoptic bronchoscope passed through the lumen of a PLMA.

**Key words:** Difficult airway, Escobar syndrome, proseal laryngeal mask airway

## INTRODUCTION

Escobar syndrome (multiple pterygium syndrome) is a rare autosomal recessive disorder.[1] Most of these patients belong to paediatric age group, with their inherent anaesthetic concerns. Optimal airway management plan is essential as these children may have cleft palate, ankyloglossia, micrognathia, restricted mouth opening, flexion contractures over neck and vertebral anomalies.[2-4] The disease is progressive, with possibility of spine deformity along with restrictive lung disease.[5] Very limited number of cases of Escobar syndrome have been reported in the literature.[1,6] We present a case of a 1-year-old child in whom difficulty in airway management was encountered during general anaesthesia. To the best of our knowledge, this is the first case report of anaesthetic management of Escobar syndrome in India.

## CASE REPORT

An 1-year-old female child diagnosed with Escobar syndrome since birth, weighing 10 kg was posted for bilateral hamstring to quadriceps transfer. Pre-operative examination revealed multiple joint flexion contractures. Airway examination showed adequate mouth opening, whereas neck extension was limited due to flexion contracture over neck. Systemic and haematological examinations were unremarkable. The lateral X-ray of the neck showed that there was no cervical spine fusion. The child also had atrial septal defect at birth, which was now closed, with normal electrocardiography (ECG) and echocardiography findings. After adequate nil per oral, patient was shifted to the operation theatre (OT) without premedication. Difficult airway cart was already kept ready in the OT. Preinduction monitors were five lead ECG, pulse oximetry and non-invasive blood pressure. Child was induced by inhalation induction with 6-8% sevoflurane and 100% oxygen. After adequate depth of anaesthesia, intravenous (IV) cannulation was carried out in the right upper limb. After checking ability to mask ventilate, 0.1 mg IV glycopyrrolate, 10 μg IV fentanyl, 5 mg IV propofol were given to increase depth of anaesthesia. Child was paralyzed with 5 mg IV atracurium, and mask ventilation was carried out with 2-3% sevoflurane and oxygen. After 3 min, intubation was attempted with conventional laryngoscopy. Intubation was unsuccessful twice wherein only the

**How to cite this article:** Mathew S, Chaudhuri S, Arun Kumar HD, Joseph TT. Airway management in Escobar syndrome: A formidable challenge. Indian J Anaesth 2013;57:603-5.

tip of epiglottis could be visualized even after optimal external laryngeal manipulation. Further attempts at conventional laryngoscopy were abandoned considering possibility of airway trauma and edema. Patient's saturation was maintained around 96-98% during this period. A size 1.5 proseal laryngeal mask airway (PLMA) was secured in place and capnogram was obtained. Ryles tube was inserted through the gastric channel to deflate the stomach. Even though there was optimal chest expansion, it was accompanied by an increase in end tidal $CO_2$ values to about 55-60 mmHg after 5 min of controlled ventilation. Furthermore, as the surgery was expected to last about 3 h, we decided to proceed with PLMA-assisted fibreoptic guided endotracheal intubation. A 4.0 mm internal diameter uncuffed portex endotracheal tube (ETT) was railroaded over the bronchoscope of 2.4 mm outer diameter and then the bronchoscope was introduced through the lumen of PLMA into the trachea. The ETT was then advanced through the lumen of the PLMA into the trachea and tip position was confirmed above the carina by visualization through bronchoscope. Equal bilateral air entry was confirmed by auscultation. The PLMA with ETT *in situ* was fixed as a single unit [Figure 1]. Anaesthesia was maintained with 66% nitrous oxide, 33% oxygen and sevoflurane 1.5-2%. With adequate ventilation, the $ETCO_2$ of 35-37 mmHg was maintained throughout surgery. Caudal analgesia was given after careful positioning with 10 mL of 0.2% ropivacaine and 10 μg clonidine. Surgery was carried out in supine position with application of tourniquet. Rest of the intraoperative period was uneventful. At the end of the procedure, child was reversed, and when she was fully awake and breathing adequately, the PLMA and ETT were removed as a single unit after oral and gastric suctioning. Child was shifted to a high dependency unit for monitoring.

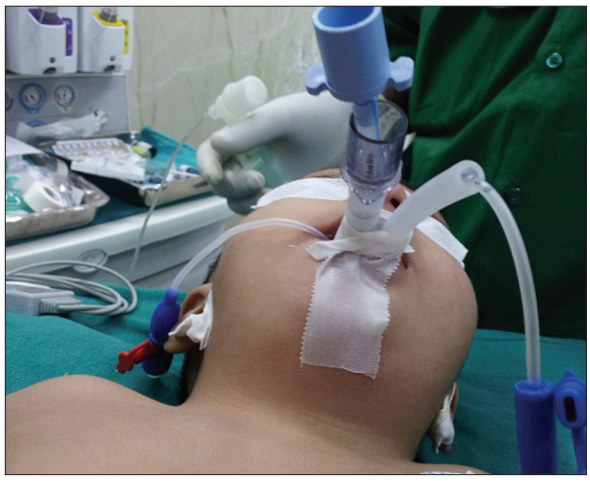

**Figure 1:** The proseal laryngeal mask airway with endotracheal tube *in situ* in the child. The neck contracture can be seen

## DISCUSSION

Children with Escobar syndrome pose multiple challenges for the anaesthesiologist. Pre-operatively, anaesthesiologist should look for evidence of micrognathia, ankyloglossia, restriction in mouth opening, webbing of neck and cleft palate to plan for optimal airway management as these can lead to difficult mask ventilation. IV cannulation may be difficult in infants with this syndrome due to the multiple contractures. Cervical spine stiffness and fusion may be present, so neck X-ray is essential.[7]

Mouth opening appeared to be adequate in our patient and inspite of limitation of neck extension, we thought that conventional laryngoscopy and intubation would be possible in the absence of cervical spine fusion.

A critical aspect of anaesthetic management in Escobar syndrome involves evaluation and sagacity in managing the paediatric difficult airway, for which various options are available.

Kuzma *et al*.[1] had used laryngeal mask airway (LMA) assisted fibreoptic guided intubation for a child with Escobar syndrome after awake fibreoptic guided intubation was unsuccessful. The authors had concluded that ETT with LMA provides a secure airway with LMA as a backup in case of ETT dislodgement.

ETT with PLMA may be more secure because of the unique cuff design. Jansen and Johnston proposed the Shikani optical Stylet as a valuable airway tool in the management such cases of difficult paediatric airway.[8] The narrow diameter of the Shikani Stylet enables it to be introduced through a restricted mouth opening and its malleability permits adaptation to unusual anatomical structures.[8]

Video laryngoscopy is an attractive alternative as it provides an all-inclusive high resolution view of the paediatric difficult airway.[9] Airtraq optical laryngoscope and GlideScope have been used for safe and successful intubation in paediatric patients with the craniofacial anomalies.[9,10] The combined use of Airtraq and fibreoptic bronchoscope (FOB) after failed tracheal intubation in grossly distorted airway has also been described, though not in paediatric age group.[11] Airtraq provides an unobstructed airway for the FOB

so that its tip can be placed near the glottis, while the FOB negotiates the sharp angle between the tip of the ETT and the glottis.[11]

We must be aware that intubation in Escobar syndrome becomes more difficult as the age of the child advances, due to increased deformity of airway by the pterygia. This is unique in Escobar syndrome compared to other congenital syndromes where management of airway becomes simpler as the child grows.[1] These children may also have pulmonary hypoplasia, kyphoscoliosis and be at risk of malignant hyperthermia.[5,12,13] Thus, anaesthesiologist should be aware of all the challenges including paediatric concerns while anesthetizing a child with Escobar syndrome.

## CONCLUSION

Difficult airway should always be anticipated in all cases of Escobar syndrome irrespective of physical and radiological examination findings and multiple airway management plans should be available. Airway management progressively becomes more difficult as the age of child advances, so a previous unremarkable airway management may become extremely difficult in future.

## REFERENCES


1. Kuzma PJ, Calkins MD, Kline MD, Karan SM, Matson MD. The anesthetic management of patients with multiple pterygium syndrome. Anesth Analg 1996;83:430-2.
2. Buyse ML. Birth Defects Encyclopedia. Cambridge, MA: Blackwell Scientific Publications 1990. p. 1427-8.
3. Thompson EM, Donnai D, Baraitser M, Hall CM, Pembrey ME, Fixsen J. Multiple pterygium syndrome: Evolution of the phenotype. J Med Genet 1987;24:733-49.
4. Valnicek SM, Clarke HM. Syngnathia: A report of two cases. Cleft Palate Craniofac J 1993;30:582-5.
5. Dodson CC, Boachie-Adjei O. Escobar syndrome (multiple pterygium syndrome) associated with thoracic kyphoscoliosis, lordoscoliosis, and severe restrictive lung disease: A case report. HSS J 2005;1:35-9.
6. Amalnath DS, Subrahmanyam DK, Sridhar S, Dutta TK. Escobar syndrome in three male patients of same family. Indian J Hum Genet 2011;17:22-5.
7. Arpaci AH, Bozkirli F, Konuk O. Anesthesic management for escobar syndrome: Case report. Case Rep Med 2011;2011:515719.
8. Jansen AH, Johnston G. The Shikani Optical Stylet: A useful adjunct to airway management in a neonate with popliteal pterygium syndrome. Paediatr Anaesth 2008;18:188-90.
9. Ramesh S, Jayanthi R, Archana SR. Paediatric airway management: What is new? Indian J Anaesth 2012;56:448-53.
10. Xue FS, Tian M, Liao X, Xu YC. Safe and successful intubation using the GlideScope videolaryngoscope in children with craniofacial anomalies. Plast Reconstr Surg 2009;123:1127-9.
11. Gómez-Ríos MA, Nieto Serradilla L. Combined use of an Airtraq® optical laryngoscope, Airtraq video camera, Airtraq wireless monitor, and a fibreoptic bronchoscope after failed tracheal intubation. Can J Anaesth 2011;58:411-2.
12. Gericke GS. Fragile collagen and the lethal multiple pterygium syndrome: Does heat stress play a role? Am J Med Genet 1991;38:630-3.
13. Robinson LK, O'Brien NC, Puckett MC, Cox MA. Multiple pterygium syndrome: A case complicated by malignant hyperthermia. Clin Genet 1987;32:5-9.


**Source of Support: Nil, Conflict of Interest: None declared**