Successful difficult airway management in a child with Hecht-Beals syndrome

Sir,

Hecht–Beals syndrome is a rare autosomal dominant inherited disorder of connective tissue. Children are born with a characteristic clenched position of hand, foot deformity, multiple joint contractures, arachnodactyly, kyphoscoliosis, limited mandible excursion and restricted mouth opening. Airway management may be challenging in these children, and various techniques like laryngoscopy, anterograde fiberoptic guided intubation, retrograde fiberoptic guided intubation, and elective tracheostomy have been reported. We put forward the use of tracheal tube introducer through laryngeal mask airway as an incomplex technique, keeping fiberoptic bronchoscope as standby for tracheal intubation in these children with restricted mouth opening.

A 4-year-old, 15-kg female child with Hecht–Beals syndrome having multiple joint contracture of the foot was posted for contracture release in prone position. Physical examination revealed contracture deformity of both upper and lower limbs, with adduction and inversion deformity of foot. Further genetic workup revealed her as a case of Hecht–Beals syndrome (FBN2 mutation). Airway examination showed small mouth opening (1.8 cm) and multiple missing teeth with normal neck movements [Figure 1]. After adequate fasting, she was shifted to the operating room.

The child was induced with 8% sevoflurane and 100% oxygen at the flow of 6 L. Standard monitoring of pulse oximetry, electrocardiography, and non-invasive blood pressure were established upon induction and a 22-G intravenous access was secured. Sevoflurane concentration was reduced to 5% and the child was allowed to breathe spontaneously. With small mouth opening, visualisation of glottis was difficult using conventional laryngoscopy and further attempts for achieving endotracheal intubation with Miller blade also failed. A classic laryngeal mask airway size 2 was inserted and adequate ventilation was achieved. A 10 CH tracheal tube introducer tip facing anteriorly was passed through the laryngeal mask airway into the trachea. The laryngeal mask airway was removed and a five millimeter internal diameter uncuffed endotracheal tube was railroaded over the tracheal tube introducer. After confirming the endotracheal placement of the tube with end-tidal carbon dioxide and auscultation for bilateral equal air entry, atracurium was administered. Caudal block

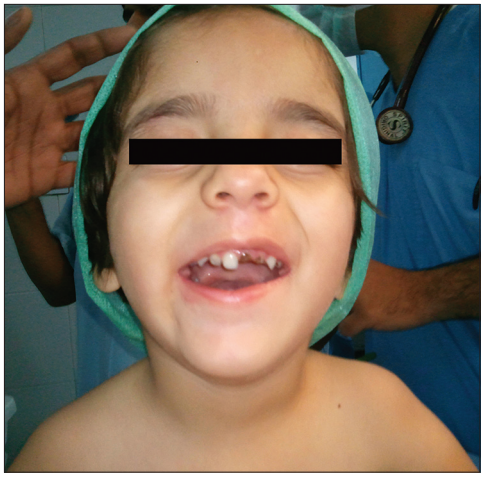

Figure 1: Child with Hecht–Beals syndrome having limited mouth opening

was achieved with 8 ml of 0.25% bupivacaine. Surgery was done in prone position. The intraoperative events were uneventful and the child was extubated when fully awake at the end of the surgery.

Hecht–Beals syndrome is a relatively rare autosomal dominant inherited single gene disorder caused due to mutation in fibrillin 2 gene (FBN2) in chromosome 5q23, affecting the connective tissue. Scoliosis, kyphosis and thoracic cage abnormalities may lead to severe restrictive lung disease. The cardiac anomalies include aortic root dilatation, aortic aneurysms, mitral valve prolapse, mitral regurgitations and ventricular septal defects. The ocular manifestations may include ectopia lentis, keratoconus and glaucomatous optic disc cupping. Trismus is due to hyperplasia of coronoid process of mandible, and tympanomandibular joint is usually normal in these children. Our patient had pseudotrismus with small mouth opening. Airway management may be difficult due to the limited mouth opening and laryngoscopy may be impossible in these patients.[1] Further attempts in laryngoscopy may result in severe airway trauma leading to complications. The technique of intubating patients with difficult airway by blindly passing bougie through laryngeal mask airway has been described with variable success rates.[2,3] Recent studies suggest that using the tracheal tube introducer with the tip facing anteriorly and rotation of the introducer may increase the success rates up to 50–60%.[4] We acclaim that this technique is unsophisticated and can be used safely in these patients with limited mouth opening, keeping advanced techniques like paediatric fiberoptic bronchoscopy as standby.

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