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Dr. Divya SS
Postgraduate, Department of Anaesthesiology, Lokmanya Tilak Municipal Medical College, Sion, Mumbai, Maharashtra, India

Dr. Shailendra Dethe
Associate Professor, Department of Anaesthesiology, Lokmanya Tilak Municipal Medical College, Sion, Mumbai, Maharashtra, India

Corresponding Author:
Dr. Divya S S
Postgraduate, Department of Anaesthesiology, Lokmanya Tilak Municipal Medical College, Sion, Mumbai, Maharashtra, India

Successful airway management in a child with Edward's syndrome: A case report

Dr. Divya SS and Dr. Shailendra Dethe

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Abstract
The airway management of syndromic pediatric patients is an ongoing challenge for the anesthesiologist. The craniofacial abnormalities and multiple system anomalies in Edward syndrome children require special consideration during the perioperative period. We hereby report the successful airway management of an 8-year-old male child with Edward's syndrome posted for laparoscopic orchidopexy.

Keywords: Difficult airway management, Edward's syndrome, anaesthesia

Introduction
Edwards' syndrome, a/k/a trisomy 18, is a genetic disorder caused by the presence of an extra 18th chromosome, named after John H. Edwards, who first described the syndrome in 1960. It is the second most common trisomy after trisomy 21 (Downs syndrome). Death often occurs in the first year of life- severe heart defects, central apnea, aspiration, respiratory failure, and upper airway obstruction- in more than 90% of patients. The usual manifestations of this syndrome are dysmorphic facies, microcephaly, flexion finger deformity, and rocker- bottom feet, delayed psychomotor development, and mental retardation. It also affects multiple organs- neurological, cardiac, pulmonary, gastrointestinal, and musculoskeletal manifestations. More than 90% of infants have cardiac malformations – VSD, ASD, PDA, TOF, and coarctation of aorta. Despite the availability of modern sophisticated techniques, the airway management of a syndromic child who has a dysmorphic face remains challenging for anesthesiologists. The purpose of this case report is to share our experience of successful airway management in an 8-year-old child with Edward syndrome scheduled for elective surgery.

Case Report
An 8-year-old male child weighing 20 kg, a full-term baby born by LSCS with a birth weight of 2.5kg, presented with intermittent pain over the bilateral inguinal region for 6 months with undescended testis and was posted for laparoscopic orchidopexy. The patient had a history of ARM repair at 2 days of age under general anaesthesia in 2014. There was no history suggestive of postoperative mechanical ventilation and ICU admission Also, he had ASD, micropenis, bilateral undescended testis, syndactyly, and dysmorphic facies at birth. The child was diagnosed as a case of Edward's syndrome at birth. Mental development and developmental milestones were delayed. On general physical examination, the child was conscious oriented comfortable while examination. He had dysmorphic facies such as a flat nose, protruding chin, a high-arched palate, low-set ears, and overlapping fingers on both hands. The cardiovascular system and respiratory system examination were normal. On airway examination – the child had an adequate mouth opening with inter incisor gap of more than 3 finger breadth and modified MPC grade 2, Thromental distance (TMD) > 6cm. Neck movements were adequate. Routine blood investigations were within normal limits. USG abdomen was suggestive of bilateral undescended testis. Chest x-ray, ECG, and Echocardiography revealed no abnormal findings.

Fig 1: Syndromic child with dysmorphic facies

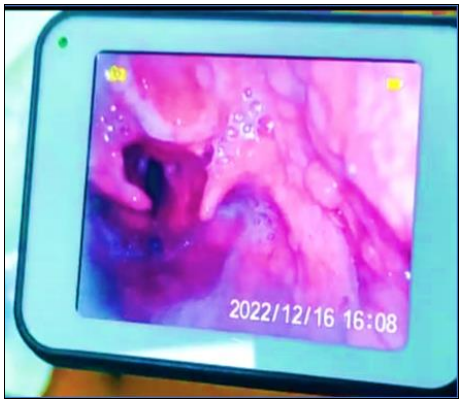

Fig 2: Videolaryngoscopic view – showing bifid epiglottis

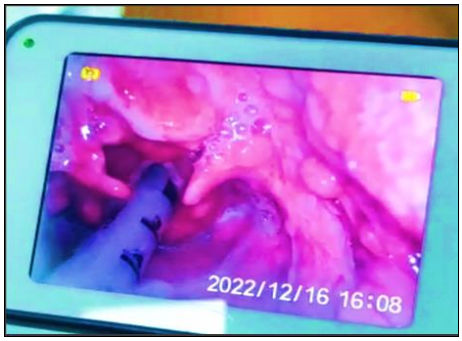

Fig 3: Tracheal intubation with 4mm microcuffed ET tube

The NBM status of the child was confirmed. Informed written consent was obtained from parents. The child was taken inside the operating theatre. A patent intravenous angiocath was in situ. ASA standard noninvasive monitor was set up which displays ECG, heart rate, noninvasive blood pressure, oxygen saturation, and program. Since the child had dysmorphic facies, we ensured that the pediatric difficult airway cart was ready. All the anesthetic drugs and emergency drugs were checked. premedication was given with glycopyrrolate 0.004mg/kg and fentanyl 1mcg/kg intravenously. The child was pre-oxygenated with 100% oxygen for 3 minutes and anesthesia was induced with propofol 3mg/kg intravenously slowly in graded doses until the loss of consciousness and centralization of the pupil. After assessing the adequacy of ventilation, an injection of atracurium 0.75mg/kg iv was given. The patient was ventilated with oxygen, air, and sevoflurane for 3 minutes. Conventional direct laryngoscopy was attempted using Macintosh blade no.3, the epiglottis was visible, but of abnormal anatomy, there was a cleft on the middle part of the epiglottis with the round structure (?swelling) on the posterior part of the tongue protruding into the cleft. So we aborted the laryngoscopy and started mask ventilation with 100% oxygen. Then the senior person was taken over and proceeded with a video laryngoscope which allowed visualization of the glottis (POGO score > 50%). Still, the trachea could not be intubated with a 5.5 mm uncuffed ET tube, which was appropriate for this age group. After two attempts and downsizing the ETT, the patient was successfully intubated with a 4mm micro cuffed endotracheal tube. Endotracheal intubation was confirmed by 5-point auscultation and square wave capnography. Anaesthesia was maintained with isoflurane, O2, air, and intermittent atracurium in a closed circuit with controlled mechanical ventilation. Analgesia was achieved with fentanyl 0.5mcg/kg and paracetamol 15mg/kg. The intraoperative period was uneventful. At the end of the surgery, we did an ENT evaluation before the reversal of the neuromuscular blockade. First, an airway assessment was done with a video laryngoscope followed by FOB. There was no evidence of subglottic stenosis or intraluminal narrowing due to laryngeal mass. ENT surgeons opined that the swelling at the base of the epiglottis was the tongue itself, prolapsing back into the defect on the epiglottis. Also, the tracheal luminal diameter was significantly reduced for his age. They suggested further evaluation of the airway anatomy with MRI postoperatively. After the reversal of residual neuromuscular blockade with intravenous glycopyrrolate 0.008mg/kg and neostigmine 0.06mg/kg, the trachea was extubated after confirming adequate minute ventilation on C-PAP PSV mode. The postoperative period was uneventful. He didn't have any signs of airway obstruction -stridor /grunting. The patient was given nebulization with salbutamol/budesonide, IV steroids, and analgesics, and monitored for 24 hours.

Discussion

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. The airway management of such patients is challenging. Effective airway management includes anticipating and planning for problems. Preoperative evaluation for congenital heart disease, other systemic abnormalities, difficulty in mask ventilation, and difficult endotracheal intubation is important. We anticipated a difficult airway in our patient because of the syndromic child with dysmorphic facies. Airway abnormalities frequently observed in Edward syndrome include micro/retrognathia, microstomia, cleft lip/palate, tracheomalacia, laryngomalacia, pulmonary hypoplasia, etc., which was not present in this child except high arched palate, protruding chin, and flat nose. In our case, intubation was difficult even though visualization of the glottis was good; adequate sized ETT (5.5,4,4.5mm) could not be passed due to the smaller internal diameter of the trachea. Finally, the trachea was intubated with a 4mm micro-cuffed ET tube which was smaller for his age. Airway assessment using video laryngoscope followed by FOB concluded that tracheal luminal diameter is smaller for his age. The normal tracheal lumen for an 8 yr old child should be approx. 9-10mm, which was significantly reduced in this child. According to the age-based formula, ID (mm) = [age in years + 16]/4 for the calculation of ETT size, the expected

tube size of this child was six uncuffed or 5.5 cuffed tubes since we were ready with smaller-sized ET tubes that could successfully intubate the child immediately without any complications. There are only a few reported cases in the literature on the anesthetic management of patients with Edwards syndrome, and as such, no standard anesthetic protocol exists. In addition, although specific airway management difficulties such as 'cannot intubate, cannot ventilate' situations were not reported, craniofacial conditions can affect intubation and airway management.

Conclusion

Airway management in syndromic children could be problematic and lead to catastrophes. Therefore, meticulous attention is required at each step. Formulating proper plans for managing difficult airways helps in successful tracheal intubation and prevents complications.

Conflict of Interest

Not available

Financial Support

Not available

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