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Case Report Open Access

General Anesthesia Preceded by Awake-Trial of LMA in a Child with Freeman-Sheldon Syndrome

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Abstract

We report the anesthetic management of a five-year-old girl child with Freeman-Sheldon syndrome undergoing surgical correction of the restricted mouth opening and the deformities of the feet. The anticipated difficult airway (mouth opening of 12 mm, MallampatiIV) was safely and successfully managed using an "awake-trial" of LMA before the actual LMA insertion under general anesthesia. Subsequent tracheal intubation through the LMA was done using a Desjardin-gallstone-forceps as a stabilization device. Perioperative period was uneventful with the complete avoidance of halogenated inhalational anesthetic agents, suxamethonium, metoclopramide, droperidol, and neuromuscular blocking agents.

Keywords: Freeman-Sheldon syndrome; Anesthetic management; Awake-trial of LMA

Introduction

Freeman-Sheldon Syndrome (FSS), or distal arthrogryposis type 2A, is a rare congenital myopathy and dysplasia characterized by multiple contractures, abnormalities of the head and face, defective development of the hands and feet and skeletal malformations. The facial muscle contracture produces the typical "whistling face" appearance. Anesthetic issues include difficult intravenous access, difficult airway, postoperative pulmonary complications, and possibly malignant hyperthermia and unpredictable responses to neuromuscular blocking agents [1].

Case Report

Written informed consent was obtained from the parents of the child before the conduct of the anesthesia related perioperative procedures. The case report was approved for scientific publication by the Institute Ethics Committee of the Medical College Hospital, Kolkata. Written informed consent was also obtained from the parents for publication of the selected photographs of the child undergoing the anesthesia related perioperative procedures in scientific journals.

The child was five-years-age, female, body weight 13 kg, height 115 cm, with a normal level of intellect (Figure 1). She was ASA physical status III on preoperative clinical assessment. Airway assessment revealed a mouth opening of 12 mm, Mallampati IV (Figure 2), normal thyromental distance and normal neck movement, indicative of anticipated difficult tracheal intubation by direct laryngoscopy under general anesthesia. The child was unable to co-operate for awake-flexible-fiberscopic tracheal intubation. One of the authors (JK) suggested the "awake-trial" of a LMA Unique™ size 2, semi-inflated (with 5 mL air), dipped in a sugar-candy syrup at room temperature. The child readily accepted to take it into her mouth just beyond the largest diameter of the LMA cuff, despite the reduced mouth opening of 12 mm (Figure 3). It was therefore confirmed preoperatively that it would be safe to induce general anesthesia in the child as the airway can easily be secured with the LMA Unique™ size 2.

Figure 1: Child with Freeman-Sheldon Syndrome.

The child was fasted for 6 hours, and premedicated with syrup triclofos 900 mg (9 mL) and oral-dispersable tablet lansoprazole 7.5 mg on the morning of surgery. EMLA™ cream with occlusive Tegaderm™ dressing was applied on a selected suitable forearm vein one hour before the intravenous cannulation.Anesthesia was induced with intravenous injections of propofol 40 mg and fentanyl 30 mcg following oxygen insufflation by a face mask held 30 cm above the face of the child. Mask ventilation (with 50% nitrous oxide in oxygen)

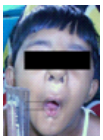

Figure 2: Airway assessment revealing mouth opening of 12 mm and Mallampati IV.

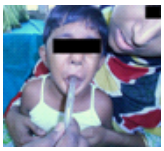

Figure 3: Awake-trial of a semi-inflated LMA Unique™ dipped in sugar candy syrup.

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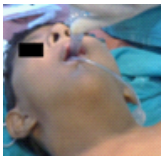

Figure 4: Successful placement of the LMA Unique™ under general anesthesia.

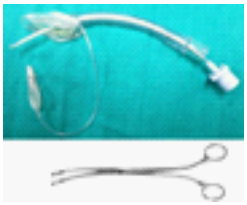

Figure 5: *In vitro* trial of the tracheal tube through the LMA Unique™.

was easy after the induction of general anesthesia. We confirmed jaw relaxation as well as loss of eyelash reflex and loss of verbal contact to determine the appropriate depth of anesthesia before attempting to insert the LMA. However, we found no improvement in the mouth opening even after jaw relaxation due to the congenital anatomical anomaly. A lubricated semi-inflated (with 5 mL air) LMA Unique™ size 2 was easily inserted inside the pharynx by the anesthetist standing at the head-end of the patient, assisted by an anesthetist applying jaw thrust, facing the first anesthetist, standing by the side of the patient. The LMA cuff was further inflated with 5 mL of air. Correct placement of the LMA was confirmed with capnography and chest auscultation on mechanical ventilation (Figure 4).

General anesthesia was maintained with 60% nitrous oxide in oxygen with intravenous propofol infusion (8 mg/kg/h for first 10 minutes, and then 6 mg/kg/h till the end of surgery) on volume control continuous mandatory ventilation (VC-CMV). Intraoperative analgesia was maintained with diclofenac rectal suppository 25 mg and incremental intravenous fentanyl bolus of 25 mcg every hour. Intraoperative monitoring included continuous ECG, pulse oximetry, capnography, temperature, airway pressure, exhaled tidal volume, exhaled minute ventilation, and automated non-invasive blood pressure at 5 minute intervals. Forced air warmer was used to maintain normothermia.

Since the patient was to undergo corrective surgery of the mouth, tracheal intubation was required. The easy passage of a size 4.0 mm internal diameter lubricated tracheal tube through the size 2 LMA Unique™ was confirmed *in vitro* before the anesthesia procedure (Figure 5). A lubricated non-cuffed-PVC-tracheal-tube of size 4 mm internal diameter was inserted through the LMA Unique™ until its connector-end was nearly inside the connector of the LMA. The correct placement of the tracheal tube was confirmed by capnography and chest auscultation on mechanical ventilation. The tracheal tube's connector was removed and the tracheal tube held steady in position by a Desjardin-gall-stone-forceps (small jaw, 23 cm length), and the LMA pulled out over the Desjardin-gall-stone-forceps. When the LMA was just out of the oral cavity the tracheal tube was grasped and kept steady at the lips of the patient. The grasp of the Desjardin's forceps was then released from the tracheal tube and the LMA taken out completely. The tracheal tube's connector was once again re-attached and correct placement of the tracheal tube was confirmed by capnography and

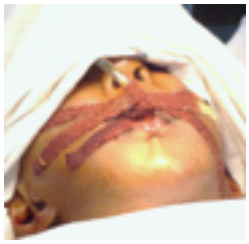

Figure 6: Successful placement of the tracheal tube.

chest auscultation on mechanical ventilation, and secured with adhesive tape (Figure 6). Throat packing was done with a saline soaked throat-pack placed on both sides of the tracheal tube in the pharynx, by blind palpation method using the index finger of the anesthetist facing the patient from the foot-end.

Intraoperative period was uneventful. On completion of the surgery, nitrous oxide and propofol infusion were stopped and throat pack removed after oropharyngeal suction. The child was extubated in recovery position on return of protective airway reflexes, adequate spontaneous minute ventilation with normal $ETCO_2$ and SpO_2, temperature, blood pressure and pulse rate being within the normal range. The postoperative period till the hospital discharge was also uneventful.

Discussion

Spinal anesthesia and combined spinal-epidural anesthesia have been described for lower limb surgeries in older children with FSS to avoid potential airway complications from anticipated difficult tracheal intubation. Airway management using awake flexible-fiberoptic bronchoscopy has been reported in FSS [2-4]. Airway management using a folded laryngeal mask airway or a non-folded laryngeal mask airway under general anesthesia has also been reported. In one case report, both tracheal intubation by direct laryngoscopy as well as the insertion of a laryngeal mask airway under general anesthesia failed due to microstomia. The child was subsequently salvaged by emergency naso-tracheal-fiberoptic-bronchoscopic intubation via a fiberoptic mask while ventilating the lungs [5-7].

In our case, the awake-trial of a semi-inflated LMA Unique™ predicted the success of LMA insertion under general anesthesia with a mouth opening as small as 12 mm, thereby increasing patient safety. The awake-trial of LMA was to test whether the largest cross-sectional diameter of the deflated LMA would pass the narrowest part of the upper airway, in this case, the narrow mouth opening. As such, the LMA was not seated in the hypopharynx during the awake-trial, which could have elicited a gag reflex and frightened the child. Also, tracheal intubation through the LMA Unique™ under general anesthesia had been successfully done with commonly available and low-cost equipments of the operating room.

The success of flexible-fiberscopic-bronchoscopic tracheal intubation in awake or sedated state depends on the co-operation of the child and the skill of the bronchoscopist, both of which are highly variable. Additionally, there are risks of hypoxia from over-sedation and partial airway obstruction. Our technique can be used as a safe and low-cost-replacement of flexible-fiberscopic-bronchoscopic tracheal intubation in selected patients with microstomia.In the case of possible failure of LMA insertion following the induction of general anesthesia, we had a backup plan of continuing with face mask ventilation after inserting a 12 Fr (3.5 mm outer diameter) or a 14 Fr (4 mm outer

diameter) lubricated nasopharyngeal airway through the nose (to facilitate face mask ventilation) until the child awakened. In that case, the surgical procedure on the mouth would have been abandoned and the child would have been sent to a center in another part of the country having the facility of pediatric flexible fiberscopic tracheal intubation.

As far as the potential risk of failure of railroading the tracheal tube through the LMA was concerned, we had two lubricated "Cook Airway Exchange Catheters (WILLIAM A. COOK AUSTRALIA PTY. LTD. 95 Brandl Street, Eight Mile Plains Brisbane, QLD 4113, Australia)". The two sizes we kept ready were one 8 Fr 45 cm (for 3 mm or larger tracheal tubes) and one 11Fr 83 cm (for 4 mm or larger tracheal tubes) ready to be passed through the well inserted LMA. Once the lubricated Cook Airway Exchange Catheter is in place, the "Rapi-fit" adapter (the 15 mm connector that comes with the catheter) of the Cook Airway Exchange Catheter will be connected and the patient ventilated. Then we would have railroaded the tracheal tube over the Cook Airway Exchange Catheter into the trachea.

A cuffed tracheal tube would have been better in our case as it offers better airway seal than a pharyngeal pack. But we used a non-cuffed tracheal tube due to the potential problem of taking out the LMA after passing a cuffed tracheal tube with the trailing pilot balloon. As predicted from the approximate formula ((age/4)+4 mm internal diameter), a 4.5 mm or 5 mm tracheal tube seemed to be appropriate for the five year old child. But preoperative physical examination revealed that the child had significant growth stunting (body weight 13 kg) and the width of her fifth finger nail was about 4 mm only. So we planned to use a 4 mm non-cuffed tube. In fact, we did not encounter any problem with peri-tubal air leakage or inadequate ventilation during the surgical procedure.

Patients with FSS are at increased risk of postoperative pulmonary complications (due to underlying myopathy), neuroleptic malignant syndrome (NMS), unpredictable responses to neuromuscular blocking agents, and possibly malignant hyperthermia (MH) [1,8]. So it was logical to avoid droperidol and metoclopramide (to avoid precipitation of NMH); halogenated inhalation agents and suxamethonium (to avoid triggering of MH); neuromuscular blocking agents (avoidance of unpredictable muscle paralysis and possible persistent postoperative muscle weakness contributing to postoperative pulmonary complications). Thus the perioperative anesthetic management in FSS can be uneventful with the complete avoidance of halogenated inhalational anesthetic agents, suxamethonium, metoclopramide, droperidol, and neuromuscular blocking agents as we observed in our patient.

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