Management of a case of anticipated difficult airway in a patient with Moebius syndrome

Sir,

Moebius syndrome is a rare neurological disorder with a prevalence of 0.0002%-0.002% of live births characterised by unilateral or bilateral facial paralysis and defective extra-ocular movements, secondary to congenital paresis of the facial (VII) and abducens (VI) cranial nerves. It can be associated with various craniofacial (mandibular hypoplasia, microstomia, temporomandibular joint dysfunction, cleft palate, external ear deformities) defects, limb (clubfoot) and musculoskeletal malformations.

We report to you a case of a 13-year-old boy weighing 38 kg diagnosed with Moebius syndrome who was posted for orthognathic surgery. The patient presented with hemifacial palsy, maxillary and mandibular hypoplasia with restricted mouth opening [Figure 1]. Airway examination revealed a mouth opening of 1.5 cm, thyromental distance of approximately 5 cm, restricted temporomandibular joint mobility and Mallampati Class IV.

Awake retrograde nasotracheal intubation was planned. This was the first choice as our fibre-optic scope was out of order. We did not consider the option of referring to any other centre as ours is a tertiary care post graduate (PG) teaching centre taking care of a lot of congenital facial deformities with difficult airway. The patient was counselled about the procedure of anaesthesia and found to be co-operative. The airway was anaesthetised by 2 ml 2% lignocaine with adrenaline soaked nasal pledges, 4 puffs 10% lignocaine spray of the throat, and 4 ml of 2% lignocaine to block superior laryngeal nerve and transcricoid injection. After piercing the cricothyroid membrane with an 18G Tuohy needle, a 0.035 floppy tip Terumo guidewire (Biorad Company) with a smooth flexible tip on one end, was introduced. Guidewire was advanced gently and when it appeared in the mouth, was taken out through the nose with the help of a Ryle’s tube inserted nasally. The flexometallic endotracheal tube was railroaded...
by inserting the guidewire through the Murphy’s eye which facilitated easy movement of the endotracheal tube preventing abutting of the tube against the anterior wall of trachea. Because the boy was very co-operative throughout the procedure except during the passage of endotracheal tube through posterior nares, mild sedation with intravenous inj. fentanyl 50 μg and inj. midazolam 1 mg was sufficient to sedate him well. Patient was monitored throughout the procedure with electrocardiogram(EGC), non invasive blood pressure (NIBP), peripheral oxygen saturation (SpO2) and end-tidal CO₂ (nasal). Correct placement of the endotracheal tube was confirmed with reservoir bag movements and capnograph. Our back-up plan was to manage any airway compromise with i-gel® supraglottic airway device, stabilise and call the patient for surgery at a later date when the fibreoptic scope was in function.

Awake retrograde intubation is an ideal cost-effective technique of airway management in these cases of anticipated difficult airway especially in developing countries as an alternative to awake fibre-optic intubation when unavailable. If the syndrome is associated with IXth and Xth cranial nerve involvement there may be dysphagia and retention of oral secretions.⁴ One of the advantages of retrograde intubation over awake fibre-optic intubation is its applicability when a lot of secretions in the upper airway makes difficult visualisation with fibrescope⁵,⁶ though fibre-optic intubation is still the gold standard.

Therefore, we conclude that this case emphasises the importance and the need to train ourselves in performing retrograde intubation as an alternative technique for any anticipated difficult airway.

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Conflicts of interest
There are no conflicts of interest.

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