Airway hitches and management in Maroteaux-Lamy syndrome

Dear Editor,

Maroteaux-Lamy Syndrome (mucopolysaccharidosis type VI [MPS VI]) is caused by deficiency of *N*-acetylgalactosamine-4-sulfatase resulting in accumulation of dermatan sulfate which evokes the varied spectra of clinical symptoms.[1] Dysmorphic facial features with multi-systemic involvement pose a formidable challenge to anesthetists.

We present a case of an 8-year-old female child with progressive bilateral flexion deformity of fingers, mouth breathing and snoring. She was posted for cervical (C1-C2) fixation surgery. Child had coarse facial features, macrocephaly (head circumference: 60 cm), macroglossia, short neck (thyromental distance: 4.5 cm, sternomental distance: 7 cm) and restricted neck mobility [Figure 1a]. Our plan A was C-MAC videolaryngoscopic (C-MAC®VL)-guided intubation with backup plans of fiber-optic bronchoscope (FOB)-guided intubation, retrograde intubation, and front of neck access as plans B, C, and D respectively. Consent of the mother was obtained. ASA standard monitors were attached. Continuous high flow nasal cannula (HFNC) oxygen therapy was started [Figure 1b]. Fentanyl 2mcg/kg (IV) was given and inhalational induction with sevoflurane was started. There was difficulty in mask ventilation even after

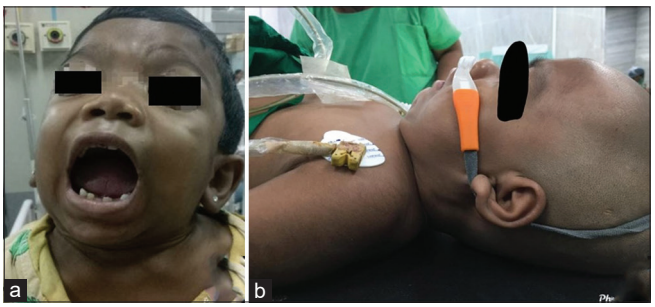

Figure 1. (a) shows facial and airway profile in this child with MPS. (b) shows continuous HFNC therapy

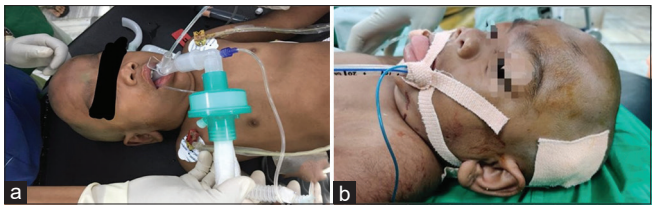

Figure 2. (a) Supreme LMA inserted to facilitate ventilation before endotracheal intubation. (b) Edematous tongue and lips noted when patient turned supine after surgery

oral airway insertion. So, immediate insertion of supreme laryngeal mask airway (LMA) size-2 enabled adequate ventilation [Figure 2a]. Injection rocuronium 1 mg/kg (IV) was administered. LMA was removed and endotracheal intubation was performed using C-MAC®VL at second attempt using bougie. Apneic oxygenation with HFNC and manual in-line stabilization was ensured until endotracheal intubation was achieved. When the patient was turned from prone to supine position after surgery, we noticed edema of the lips and tongue [Figure 2b]. Videolaryngoscopy revealed edema of posterior commissure which resolved with intravenous steroids and then extubated 24 hours later in ICU.

Maroteaux–Lamy syndrome is a type of mucopolysaccharidosis characterized by learning difficulties, mental retardation, coarse facial features, frontal bossing, depressed nasal bridge, macroglossia, adeno-tonsillar hypertrophy, narrowed airway, short, stiff neck and temporomandibular joints leading to difficult airway.[2,3] Multi-systemic involvement includes obstructive or, restrictive lung disease, valvular heart disease and cardiomyopathy. Instability of the atlantoaxial joint requires special care during intubation to prevent further compression of the cervical spinal cord.[4] Difficulty in maintaining adequate ventilation may be due to obstructive and restrictive lung disease. Intraoperative hemodynamic instability may occur due to underlying cardiac disease. Prone position can further aggravate existing airway narrowing as it can lead to airway edema which may pose difficulty during extubation.

In our case, we chose inhalational induction over intravenous induction as it was anticipated difficult airway. We could overcome the difficulty of mask ventilation by expeditious insertion of LMA.

C-MAC®VL-guided intubation was chosen over the fiber-optic technique as it provides fixed wide view of the glottis that aids in quick recognition of airway landmarks.[5] FOB-guided intubation would be difficult as the patient had poor comprehension and, airway collapse leads to difficulty in maneuvering and advancement. HFNC oxygen therapy prevented desaturation. Thus, from this case, we infer that in a child with mucopolysaccharidosis, inhalational induction, ventilation with LMA followed by intubation with C-Mac videolaryngoscope would be a logical method of airway management.

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Conflicts of interest

There are no conflicts of interest.

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