**Management of a difficult airway in a newborn with Moebius Syndrome**

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**Introduction**

Moebius sequence is a rare congenital syndrome with multiple deformities, including cranial nerve palsy (VII and VI), craniofacial anomalies and limb abnormalities. Less commonly, other cranial nerves may be involved as well. Obstructive and/or central sleep apneas are frequently seen. The central component is related to an underdeveloped respiratory center and the obstructive component to abnormal airway anatomy and physiology. Due to the deformities, these patients require a variety of reconstructive surgeries. The anesthetic management of patients with Moebius sequence focuses mostly on airway challenges with obstructive sleep apnea (OSA) and difficult intubation. We report a case of an infant with difficult airway, requiring special maneuvers to maintain airway access during tracheostomy procedure.

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**Case Summary**

- 3-month-old male infant born at 40 weeks with Moebius sequence history of difficult intubation required elective tracheostomy primarily due to severe obstructive apnea and secondarily difficult airway.
- On examination, he had typical Moebius characteristics with VI and VII nerve palsy, cleft palate, craniofacial deformity, micrognathia, microstomia, club feet, and hearing loss from VIII nerve palsy. He had severe OSA with an apnea-hypopnea index (AHI) 72 and oxygen desaturations at 45%. He was maintained on 0.75 L/min nasal cannula for 3 months prior to intubation. A 3.0 uncuffed endotracheal tube (ETT) was secured.
- Spontaneous ventilation was maintained with Sevoflurane and Morphine infusion. When the tracheotomy was created, a 4.0 Fr blunt-tipped ureteral catheter was inserted inside the ETT through the bronchoscopy swivel adapter while the ventilation was maintained, serving as a tube exchanger in case the tracheostomy tube placement failed and the ETT was to be advanced. The ETT then was withdrawn slowly over the catheter to allow the tracheostomy tube to enter the trachea. Easy ventilation through the tracheostomy tube was confirmed with bilateral breath sounds and the ETT was removed with the catheter. The rest of the case was uneventful and the patient was transported back to PICU in stable condition.

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**Discussion**

- Anesthetic management in Moebius sequence mostly involves the difficult airway with OSA and abnormal airway anatomy leading to the “difficult ventilation- difficult intubation” situation.
- OSA in Moebius sequence is anatomical and could be treated temporarily with nasal CPAP while awaiting the reconstructive surgery. However nasal CPAP in newborns and young infants are sometimes unsuccessful due to lack of compliance from patients and/or parents.
- Tracheostomy is indicated for severe OSA in which nasal CPAP is not effective or applicable. The most critical point during infantile tracheostomy is when the ETT is being withdrawn to allow the entrance of the tracheostomy tube. There is the risk of losing the airway when the tracheostomy tube fails to fit in place and the ETT cannot be reinserted while being partially out, especially in the presence of difficult airway. An ETT exchange catheter could help guide the ETT back into the trachea. However, there may not be small-sized catheters available to fit 3.0 or 3.5 ETT at many institutions. Blunt ureteral catheters are available in 3.0 Fr and 4.0 Fr sizes and could serve as an ETT exchange catheter while allowing ventilation through the lumen.

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**References**