Perioperative management for release of congenital alveolar synechiae in a 5 day-old term neonate with Van der Woude Syndrome

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Introduction:
Van der Woude’s syndrome is a rare genetic disorder characterized by cleft lip, cleft palate, and lower lip pits. We present a case associated with intraoral synechiae that posed a challenge for airway management.

Case Report:
A term infant was transferred to our hospital 11 hours after birth for further management of cleft palate complicated by severely limited mouth opening. Evaluation by plastic surgery revealed bilateral lip pits, cleft palate, interalveolar synechiae, and synechiae of the tongue to the rudimentary palate limiting mouth opening to 3mm. The constellation of physical symptoms was concerning for Van der Woude’s syndrome which was verified by the genetics team. A flexible fiberoptic nasopharyngeal scope performed bedside by the otolaryngologist confirmed the oral cavity findings and revealed normal lower airway anatomy of the larynx and trachea. On day of life five, the patient presented from the NICU for exam under anesthesia and release of congenital alveolar synechiae. He did not have any respiratory distress but was unable to feed due to his limited mouth opening. This presented a challenging airway management for the anesthesia team. General anesthesia was induced with sevoflurane while the patient maintained spontaneous ventilation. A size 1 LMA was then placed. The plastic surgery team then released the alveolar synechiae resulting in significantly improved mouth opening. The LMA was removed at the end of the case, and the patient transported back to the NICU. The patient did well and was discharged home. He did return to our hospital at age 11 months for cleft palate repair. At that time, he was intubated without difficulty. Post-operative follow-up reveals that he is doing well without feeding or respiratory difficulties.

Signs of Van der Woude’s Syndrome1,2:
- Cleft lip
- Cleft palate
- Cleft uvula
- Lower lip pits
- Accessory salivary glands
- Hypodontia
- Syngnathia
- Oral synechiae

Discussion:
Van der Woude’s syndrome in association with intraoral synechiae is concerning for airway management. In cases of respiratory distress, emergent surgery must be performed. However, even without respiratory distress early treatment is still indicated to allow for oral feeding and to prevent temporomandibular joint ankylosis. As a result, these infants present at an early age for surgical repair. Limited mouth opening can prevent oral intubation with direct or video laryngoscopy. Case reports have described using nasal fiberoptic intubation.3 This case presents an alternative approach using an LMA and spontaneous ventilation to successfully manage the airway until surgical release improved mouth opening.

References: