

MANAGING TBAO IN ROBIN SEQUENCE WITH CONCOMITANT MULTI-LEVEL AIRWAY DISEASE

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Background: Pierre Robin sequence (PRS) is described as a triad of micrognathia, glossoptosis, and tongue-based airway obstruction (TBAO). When concomitant airway anomalies are present, mandibular distraction alone is often unsuccessful at alleviating the obstruction, mandating the need for tracheostomy. We present the first-reported case of PRS with concomitant complete tracheal rings and highlight the importance of collaborative surgical efforts to avoid tracheostomy.

Materials/Methods: A 9-day-old male presented for evaluation of a difficult airway following failed intubation. Micrognathia, glossoptosis, and cleft palate were found on physical examination, and dynamic airway CT imaging demonstrated complete tracheal rings. To correct all underlying airway pathologies, a slide tracheoplasty followed by mandibular distraction osteogenesis (MDO) was performed. Though transient respiratory events occurred distantly following surgery, with conservative management, the patient was eventually extubated and has since been stable on room air.

Results: Tracheostomy is a major concern when managing TBAO in patients with PRS and depends on timing and severity. The long-term sequelae of a tracheostomy include tracheal stenosis, infections, and sudden death. Our patient's individualized management approach was devised by six teams, including plastic surgery, and resulted in a successful two-staged surgical approach that avoided the consequences of tracheostomy. The complexities of this patient's management highlight the importance of multidisciplinary collaboration for optimal delivery of care.

Conclusions: In the setting of severe PRS-related respiratory distress unable to be managed conservatively, securing the airway and carefully evaluating for concomitant airway anomalies are of highest priority. In the setting of multiple airway anomalies, a collaborative team mentality can decrease morbidity, improve long-term results, and lead to an alternative to the current paradigm of care.