

## **Treacher Collins Syndrome and Tracheostomy: Decannulation utilizing Mandibular Distraction Osteogenesis.**

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### **Abstract**

**INTRODUCTION:** Treacher Collins (TC) and Robin Sequence (RS) are both associated with hypoplastic, retrognathic mandible, glossoptosis, and airway obstruction. Bilateral Mandibular Distraction Osteogenesis (MDO) has been used successfully [1] in the past two decades [2] to treat severe airway obstruction and provide a way of enabling successful removal of tracheostomy in the RS population [3]. TC is associated with aberrations of the lower jaw, upper jaw and nasal airway [4] and is distinct from RS. Although in both TC and RS, airway obstructions in severe cases will necessitate surgical intervention, the different pathophysiology suggests MDO to be less successful in TC compared to the current literature on RS. The purpose of this study is to report on the clinical outcomes of tracheostomy removal utilizing MDO in the TC patient population.

**MATERIALS AND METHODS:** Between 1991 and 2010, 63 patients underwent bilateral MDO in our institute. The exclusion criteria were any diagnosis other than TC or insufficient medical records. Our cohort included 20 patients with tracheostomy (18 TC and 2 Nagar syndrome patients). MDO data, comorbidities, airway evaluation and complications were collected and analyzed.

**RESULTS:** The distraction devices used were external in 67% and internal in 33% of cases. The mean age of the first MDO was 4.97 years. The mean number of distractions was 1.42 with 46% of patients who had more than one-distraction attempted. Decannulation was accomplished in 15 patients (75%). Complications were divided into major (ankylosis, device failure) moderate and minor (pin infection, hypertrophic scar). Overall, 67% of TC patients had at least one complication with 41% having major complications.

**CONCLUSION:** Retrognathia and glossoptosis result in airway obstruction in both TCS and RS with similar respiratory difficulties and clinical findings. However these craniofacial abnormalities have a very different pathophysiology and should not be considered as equal entities in airway obstruction management and surgical outcomes. Combination of delayed primary MDO, which is done mostly for decannulation and the underlying anatomical differences lead us to believe TCS is a different entity of micrognathia, with a lower rate of surgical success and much higher rate of complications.

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