Rehabilitation of Micrognathia and Microglossia with Mandibular Distraction Osteogenesis

Erik Berg, MD and Andrew R. Scott, MD

Department of Otolaryngology – Head & Neck Surgery, Tufts Medical Center, Boston, MA

ABSTRACT

Objective: To describe an approach to surgical rehabilitation of feeding, speech, and upper airway obstruction in an infant with microglossia and micrognathia.

Methods: Case report and literature review

Results: We describe a 17-month-old child with micrognathia, microglossia, and submucous cleft palate who was initially fed via g-tube and did not require early airway interventions. As the child was attaining normal developmental milestones, the decision was made to perform bilateral mandibular distraction osteogenesis as a means of improving feeding, speech, and clinical symptoms of obstructive sleep apnea. In this report, relevant literature is also reviewed as it pertains to the rehabilitation and reconstruction of this rare craniofacial anomaly.

Conclusion: Mandibular distraction osteogenesis improved glossoptosis and created a more favorable position of the tongue within the oral cavity. This in turn allowed for an overall improvement in oral motor function facilitating removal of the feeding tube.

INTRODUCTION

Mandibular distraction osteogenesis (MDO) has become a widely-accepted treatment for micrognathia with upper airway obstruction.1,2 Indications for distraction include mandibular hypoplasia associated with a number of diagnoses including Pierre Robin Sequence, Nager Syndrome, Treacher Collins Syndrome, Goldenhar Syndrome, and Oromandibular Limb Dysgenesis among others.3 The clinical finding of isolated microglossia and micrognathia is extremely rare, with the first case description credited to a French physician de Janvry in 1718.4 Of all these reported cases, little has been reported on treatment. Within this report we present a case of isolated microglossia and micrognathia manifesting as speech delay and feeding difficulties, which was successfully treated with mandibular distraction osteogenesis.

CASE PRESENTATION

AR was born to a 21 year old mother at 40 weeks via cesarean section secondary to fetal distress and polyhydramnios. He had noticeable craniofacial abnormalities at birth and Otolaryngology consultation was obtained. Initial evaluation was notable for micrognathia and a hypoplastic, posteriorly positioned tongue. There was stertor with supine positioning. A flexible laryngoscopy confirmed the above findings and also noted mild laryngomalacia. A Feeding Team evaluation with modified barium swallow confirmed oral phase dysphagia as well as frank aspiration. A Genesius evaluation was unremarkable; ultrasound showed a normal thyroid. A gastrostomy tube was placed and he was discharged home. At 7 months old he was showing signs of speech delay as well as continued gastrostomy tube dependency. A computed tomography scan of mandible was obtained (Figure 1) and showed normal condylar growth. A sleep study ordered was noted but unremarkable. A pediatrician believed it was normal.

In an effort to improve his articulation and oral phase dysphagia, the patient underwent bilateral MDO with external distraction devices and the mandibular bodies were advanced approximately 20 mm prior to consolidation. Following a 14 day activation phase and a 7 week consolidation phase the hardware was removed (Figure 2). Postoperatively he had resolution of glossoptosis (Figure 3), improved oropharyngeal airway (Figure 4), and an improvement in mandibular projection and occlusion. (Figure 1 and 5) He feeds orally, is no longer completely gastrostomy tube dependent, and has improved speech.

DISCUSSION

The findings of isolated microglossia can be explained with an understanding of head and neck embryology. Our patient had a defect of the 3rd arch, the 3rd pharyngeal arch derivative, and a normal pharyngeal tongue, a 4th & 5th pharyngeal arch derivative. (Figure 6) The mandible is also a 1st arch derivative. Thus, our patient had an isolated 3rd arch defect.

Hypothetical causes of isolated microglossia vary from vascular anomalies, to syndromes, medications, and tongue anomalies, but none of these theories have been substantiated. Microglossia may also be associated with distal limb abnormalities.

During the work-up of isolated microglossia, it is important to check for functional thyroid tissue because the thyroid develops at the same time as the tongue during the 3rd week gestation. Thus, the mandibular condyles must be assessed, as absence of these structures is a contraindication for early MDO.

When this child was a neonate, we elected for conservative management because the patient had mild airway obstruction and an unknown neuro-developmental prognosis.

The evidence for performing MDO for feeding difficulties is also somewhat limited. Lishky et al made a case for using early MDO to resolve feeding difficulty in children with Pierre Robin Sequence. In a subset of 53 isolated, non-syndromic, Pierre Robin patients, the authors found that 14 of 14 (100%) of early MDO patients did not require a G-tube.5 Another study by Gourley et al looked at 10 infants, treated early with MDO.6 After 5 years, there was a high relapse rate from inadequate mandibular growth compared to the multistage approach which resulted in feeding difficulties after resolving patients without significant airway compromise.

Our patients did not fit into either category (Pierre Robin with airway compromise or syndromic); thus close observation seemed appropriate. When he clearly had speech delay & remained G-tube dependent at 17 months of age, MDO was performed to improve his speech and feeding.

Although MDO improved glossoptosis, this child will continue to benefit from multidisciplinary support for his isolated microglossia, with continued speech & swallow therapy and other possible interventions, to make further progress.

CONCLUSIONS

Isolated Microglossia/Micrognathia is rare and causes various feeding and speech difficulties. Treatment may include bilateral MDO to improve patient’s airway, feeding, and/or communication skills, although little evidence supports the latter two. Timing of MDO depends on severity of symptoms and MDO is only one component of many interventions in the context of a multidisciplinary rehabilitation.

REFERENCES

3. Sidman JD, Sampson D, Templeton B. Distraction osteogenesis of the mandible for airway development of the tongue. The oral (ant. 2/3rd) tongue from (orange) 1st pharyngeal arch, & pharyngeal (post. 1/3rd) tongue from (green) 2nd & 3rd pharyngeal arches (blue). 3rd & 4th pharyngeal arches.