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PURPOSE: Infants with RS have airway obstruction of varying severity that dictate their management (non-surgical vs. surgical). In our centre, all infants with RS and significant airway obstruction have sequential evaluation with oximetry in addition to timed polysomnography. The purpose of the study was to characterize, for the different treatment strategies, the evolution during the first year of life and post palatal closure.

METHODS: We reviewed all oximetry and polysomnography data accumulated in infants who presented with significant airway obstruction. Treatment consisted either of prone position (prone, n=11); use of nasopharyngeal airway (NPA, n=4); tongue-lip adhesion (TLA, n=6); or mandibular distraction osteogenesis (MDO, n=5). Desaturation index (drops in SpO2 ≥ 4%, DI4%; normal ≤ 8 events/h) and % time <90% (normal ≤ 0.2%) were used as parameters for airway obstruction on oximetry. Central apnea-hypopnea and mixed obstructive apnea-hypopnea index (MOAHI) were used from PSG.

RESULTS: On the initial oximetry (pre-treatment), DI4% and % time < 90% was higher in TLA, MDO and NPA groups than in the prone group (p<0.001). In the first week post-surgery, 3 neonates had persistence of high DI4% due to frequent brief central apnea. All infants improved over time, however, improvement was faster for infants undergoing surgery and much slower for NPA. By 3 months, there was no longer a difference between the two surgical groups and the prone group.

In infants with persistently elevated DI4% on oximetry (DI4% >10 ev/h, 4 prone, 3 TLA, 2 MDO), PSG showed predominance of brief central apnea with drops in oxygenation; MOAHI was < 5 events/h (normal or mild obstruction).

Before palatal closure, all infants except one in the NPA group (needed MDO) had normal or near-normal oximetry and polysomnography. Post palatal closure, 4 infants (36%) who had surgery in the neonatal period deteriorated; in the TLA group, 2 needed adenoidectomy, with one eventually needing MDO. In the MDO group, 1 infant needed adenoidectomy and one prolonged CPAP treatment. In that group that deteriorated post palatal closure, the MOAHI went from 4.4 events/hour (mild obstruction) to 35.8 events/h (severe obstruction).

CONCLUSION: TLA and MDO both resulted in the most rapid improvement in obstruction parameters, and the use of NPA was associated with the longest time to improve. Brief central apnea is a frequent occurrence and PSG was needed for the diagnosis. Infants treated surgically for obstruction in the neonatal period had less favorable outcome post palatal closure, likely reflecting more severe initial obstruction due to narrow retropharyngeal space or associated airway anomalies.

Tongue Lip Adhesion Outcomes and Mechanism of Obstruction in Robin Sequence

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PURPOSE: Increasingly, data shows favorable outcomes with regard to surgical airway management with the less-invasive Tongue-Lip Adhesion (TLA), and unfavorable long-term sequelae of neonatal distraction, raising questions of its necessity. This purpose of this study is to show pre- and post-op polysomnography data in children treated with tongue-lip adhesion, with and without oxygen supplementation, that demonstrate dramatically improved airway function over the first six months of life. This series describes an approach that can obviate in most cases the need for mandibular distraction, even in severe and syndromic forms of Robin sequence.

METHODS: All patients meeting clinical criteria for Pierre Robin Sequence (PRS) and who had undergone tongue-lip adhesion with pre- and post-op sleep studies were reviewed. The clinical series included 17 subjects (13 non-syndromic, 4 with Stickler syndrome). All but one were managed either by TLA alone or in combination with positioning and/or supplemental oxygen; the 17th (Stickler syndrome) subsequently underwent mandibular distraction;
an additional nonsyndromic patient underwent mandibular distraction prior to palatoplasty. None underwent tracheostomy. Most received gastrostomy to insure optimal nutrition.

RESULTS: Of the 13 nonsyndromic patients managed with TLA as definitive treatment, the mean preop apnea-hypopnea index (AHI) was 37.5 (range 21.0–71.8); the mean initial postop AHI improved to 7.7 (range 0.9–31.0). Of the four syndromic patients, the mean preop AHI was 65.5 and improved to 23.1 postop, with one failing TLA and subsequently treated with mandibular distraction and a post-distraction AHI of 1.0. By approximately six months of age the AHI among both syndromic and nonsyndromic patients had improved further to a mean of 1.8 (range 0–5.3) with the addition of oxygen in patients with initial postop AHI>10. Of those treated with oxygen in addition to TLA, all showed improvement in AHI, and all were weaned prior to palatoplasty. Overall growth was essentially normal. “Catch-up” mandibular AP growth was clinically variable, with followup varying from ten months to 11 years.

CONCLUSION: TLA provides a less-invasive and reliable treatment option for the majority of cases of PRS, including syndromic forms; mandibular distraction remains an essential surgical option but is necessary only in select cases. Furthermore, the clinical successes and failures of this protocol provide new insight in the pathophysiology of the airway problem in Robin sequence, and substantiates the theory that in most cases of newborn retrognathia functional recovery will occur with time and growth.

Mandibular Distraction and Long-Term Dental Morbidity

Presenter: Wendy Chen, MD, MS

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Mandibular distraction osteogenesis (MDO) has become a popular intervention since it was described for correcting hypoplastic mandibles 25 years ago. However, it remains a relatively recent tool, and long-term complications are unclear. This study reports dental morbidities and mandibular morphological findings in patients who have undergone MDO.

METHODS: This is a retrospective review of patients at our institution who have undergone MDO. Inclusion criteria involved patients with radiographs and orthodontic evaluation at early mixed and early permanent dental development. Data included demographic information, medical and surgical details, complications, and dental information, i.e. morphologic anomalies, missing teeth, aberrant anatomical locations, and buccal soft tissue redundancy. These findings are compared to normative controls.

RESULTS: Twenty-two patients met inclusion criteria: 19 Caucasian (86.4%); 1 Hispanic, 2 African American), 13 male (59.1%), 12 patients with diagnosed syndromes (54.5%), and 9 patients with hemifacial microsomia (40.9%). On average, age at first MDO was 7.95 years; 9 patients required ≥1 distraction, 10 patients required tracheostomy. Average age at last follow up was 15.1 years. On evaluation at early mixed dentition, only two patients had undergone MDO as an infant (<1yo) and had an image available for review. 3rd molars could not be visualized on either patient, but this is within age-appropriate dental development. Overall, of 22 mandibles at this age, all patients had some dental anomaly: there were a total of 8 missing molars (1st molar =2, 2nd molar =6; third molars not accounted for due to developmental stage), one impacted 1st molar, four ectopic 2nd molars, one ectopic 1st molar, five dysplastic molars (1st molar =4, 2nd molar =1), two missing 2nd premolars, two impacted premolars (1st premolar =1, 2nd premolar =1); one impacted canine and one impacted lateral incisor. Two patients had a V-shaped sigmoid notch. Two patients had unfavorable buccal mucosal scarring; 3 patients had anterior open bites described in their history; none had facial nerve complications reported.