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Sleep architecture in Pierre-Robin sequence: The effect of mandibular distraction osteogenesis

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ABSTRACT

Introduction: Pierre-Robin Sequence (PRS), a triad of micro/retrognathia, glossoptosis, and upper airway obstruction, usually in conjunction with a cleft palate is frequently associated with significant morbidity. Mandibular distraction osteogenesis (MDO) is an effective treatment modality to address retroglossal upper airway obstruction by increasing the anterior-posterior diameter of the infant airway. Although MDO has been shown to improve the apnea-hypopnea index (AHI) in children with PRS, the consequences of MDO on other aspects of infant sleep, including hypercapnea, hypoxia, the REM to Non-REM ratio, as well as its effect on central and mixed apneas has not been investigated with an adequate sample size.

Objective: To characterize the effect of MDO on key components of sleep architecture in infants with PRS.

Methods: Charts from 32 infants with PRS that were addressed with MDO at our tertiary-care children's hospital were retrospectively reviewed. Of these, 26 infants (57.7% male; mean age = 4.1 weeks, SD = 5.0) had pre- and post-operative polysomnograms (PSG). Paired samples t-tests were used to compare pre- and post- MDO sleep architecture mean score differences.

Results: Among the 26 infants, 73.1% demonstrated severe pre-MDO sleep apnea (AHI > 10). Several aspects of sleep architecture were found to improve post-operatively. Significant improvements were found in AHI (30.3 vs. 8.7; t = 4.1, $p < 0.001$), obstructive apneas (79.3 vs. 5.8; t = 4.0, $p < 0.001$), hypopneas (48.1 vs. 22.1; t = 2.2, $p = 0.040$), time spent below 90% SpO2 (3.9% vs. 0.7%; t = 3.3, $p = 0.003$), and lowest SpO2 nadir (75.4% vs. 82.9%; 3.4, $p = 0.002$). In addition, a marginally significant improvement was found for mixed apnea (6.3 vs. 1.6; t = 1.99, $p = 0.058$).

Conclusion: MDO improves several sleep architecture parameters in this sample of infants with PRS. Statistically significant improvement was seen in obstructive apneas, hypopneas, AHI, obstructive AHI, and several indicators of hypoxia during sleep.

1. Introduction

Pierre-Robin sequence (PRS), a triad of micro/retrognathia, glossoptosis, and upper airway obstruction was first described by the French stomatologist Pierre Robin in 1923 [1]. Although phenotypically variable, the sequence is frequently associated with a u-shaped cleft palate [2]. The mandibular hypoplasia results in glossoptosis, which in turn causes upper airway obstruction, feeding difficulties and sleep-disordered breathing (SDB) [3,4].

SDB, primarily in the form of obstructive sleep apnea (OSA), is a near universal finding in infants with PRS, with prevalence ranging between 85 and 100% [3,4]. The morbidity associated with OSA in infants has been well characterized, and includes failure to thrive, developmental and learning delay, cor pulmonale, and death [5]. Costa et al. in a large retrospective review found the mortality of PRS to be 16.7%, interestingly there were no deaths in isolated PRS infants [6].

Recent publications have advocated for routine PSG testing on

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all patients with PRS presenting with feeding difficulties, as well as those with desaturations in the prone position [7]. The objective nature of the PSG makes it an ideal tool to guide airway management and prevent OSA related morbidity [8].

Conservative approaches to PRS patients with upper airway obstruction include prone positioning, a nasopharyngeal airway (NPA), and continuous positive airway pressure (CPAP). There is mixed literature regarding efficacy and tolerability of NPA and CPAP for long-term management of infant upper airway obstruction [9,10]. Those patients with severe obstructive symptoms are less likely to tolerate long-term conservative measures, and are appropriate surgical candidates [9]. Studies have shown that only 10% of isolated PRS infants will require surgical management [11]. Surgical options for PRS upper airway obstruction include tongue-lip adhesions (TLA), mandibular distraction osteogenesis (MDO), and tracheotomy.

TLA has historically been used as an effective means to relieve upper airway obstruction and prevent tracheostomy in PRS infants [12]. The procedure involves surgically attaching the ventral tongue to the lower lip in an effort to alleviate base of tongue obstruction [10]. This technique has been largely supplanted by MDO due to difficulties with feeding, wound breakdown, and inadequate reduction in upper airway obstruction [9,10]. Numerous studies have demonstrated superiority of MDO over TLA in patients with PRS [13,14]. By augmenting the anterior-posterior dimensions of the infant airway, MDO has also recently been shown to prevent tracheostomy in patients with PRS, and to result in a successful decannulation in already tracheostomized patients [15]. While it is more effective, MDO carries inherent risks of inferior alveolar and marginal mandibular nerve injury, bony malunion/nonunion, tooth bud injury, and temporomandibular joint ankyloses [12,13].

Contrary to historical beliefs, the severity of untreated PRS-related OSA in infants may not improve with time, as evidenced by unchanged AHI, obstructive-AHI, and central sleep apneas (CSA) for up to 1 year [16]. Surgical treatment with MDO has been shown to improve the AHI in several studies [10,17]. However, the consequences of MDO on other aspects of infant sleep including hypoxia, the REM to non-REM ratio, hypercapnea, as well as its effect on central and mixed apneas have not been investigated with an adequate sample size. This study aims at determining the effect of MDO on sleep architecture in infants with PRS. We hypothesize that MDO will demonstrate significant improvements in post-operative PSG with regard to hypoxia, hypercapnea, mixed apneas, hypopneas, obstructive apneas, and AHI.

2. Methods

The charts of infants aged 0–3 months with a diagnosis of PRS who underwent MDO at the Children's Hospital of Michigan between 2008 and 2014 were retrospectively reviewed. Institutional review board approval was obtained from both Wayne State University and Detroit Medical Center prior to data collections. Exclusion criteria included age greater than 3 months, incomplete medical records, and lack of either pre or post-MDO PSG. Charts were reviewed by 2 independent Otolaryngologists with 25% of charts reviewed by both with greater than 90% inter-rater reliability. All data was stored in a HIPPA-compliant database.

All infants underwent a general bedside physical examination, sleep endoscopy, direct laryngoscopy, rigid bronchoscopy, and PSG. The sleep endoscopies involved flexible fiberoptic pharyngolaryngoscopy with attention to the airway in the supine and lateral recumbent positions, as well as with tongue protrusion and jaw thrust. All infants were treated with mandibular distraction using either KLS or Stryker devices by the same plastic and reconstructive surgery team. Post-operative PSG was obtained on all patients at the conclusion of the distraction process but prior to removal of hardware.

Data was collected retrospectively on patient age, gender, associated syndromes, pattern of upper airway obstruction, associated airway lesions, and both pre- and post-MDO PSG. FFL findings were detailed with base-of-tongue collapse in various positions as well as findings of laryngomalacia. Laryngomalacia was graded as mild, moderate, and severe in terms of supraglottic collapse by one of four pediatric otolaryngologists. Signs of reflux, as well as additional airway lesions were also documented based on physical exam findings at the time of FFL and direct laryngoscopy and bronchoscopy.

Pre- and post-MDO PSG data collection included number of obstructive, mixed, and central apneas, as well as AHI, lowest SpO2 nadir, time spent below 90% SpO2, and REM to non-REM ratio. Unfortunately, CO2 data was not available on most PSGs due to the technical difficulty of obtaining CO2 measurements in the infant population. All sleep studies were performed at the sleep laboratory at Children's Hospital of Michigan and interpreted by a board-certified Pediatric Pulmonologist certified in sleep medicine. When multiple sleep studies were available, the studies closest to the time of surgery (distraction or hardware removal) were used.

Paired samples t-tests were used to compare means of pre- and post-MDO measurements of sleep architecture. Univariate analysis was used to determine whether associated findings of laryngomalacia, bronchomalacia, reflux, or associated syndromes played a role in outcomes for MDO.

3. Results

32 charts of infants with PRS who underwent MDO were reviewed, 6 of which were excluded, due to either incomplete documentation or due to the absence of a post-operative PSG, resulting in 26 included infants (57.7% male, 42.3% female; mean age = 4.1 weeks, SD = 5.0, Range = 0.57–21 weeks) who met inclusion criteria.

Severe sleep apnea (AHI > 10) was identified in 73.1% (mean pre-MDO AHI = 30.3, SD = 27). Of note, one infant required supplemental oxygen and nasal trumpet during the pre-operative PSG due to severe obstruction. The mean number of pre-MDO hypopneas, obstructive, mixed, and central events were 48.1 (SD = 62.9), 79.3 (SD = 98.4), 6.3 (SD = 12.42), and 5.65 (SD = 11.5) respectively. Associated laryngomalacia, tracheomalacia, bronchomalacia was identified in 13 (52%), 6 (24%), and 2 (8.0%) of infants respectively. These secondary sites of airway obstruction were not associated with significant changes in post-MDO outcomes. Reflux was identified in 7 (27%) of infants, and again no statistically significant differences were identified on pre or post-MDO PSG.

Several aspects of sleep architecture were found to improve post-operatively. Significant improvements were found in AHI (30.3 vs. 8.7; $t = 4.1$, $p < 0.001$), obstructive AHI (27.7 vs. 5.4; $t = 4.8$, $p < 0.001$), obstructive apneas (79.3 vs. 5.8; $t = 4.0$, $p < 0.001$), hypopneas (48.1 vs. 22.1; $t = 2.2$, $p = 0.040$), time spent below 90% SpO2 (3.9% vs. 0.7%; $t = 3.3$, $p = 0.003$), and lowest SpO2 nadir (75.4% vs. 82.9%; 3.4, $p = 0.002$) (Fig. 1). Neither central AHI nor mixed AHI were significantly altered by MDO (1.2 vs. 3.45; $t = -1.39$, $p = 0.177$ and 6.35 vs. 1.6; $t = 1.96$, $p = 0.56$). To date, no patients have required a tracheostomy as part of their airway management.

4. Discussion

MDO was initially introduced in the 1990's for infants with hemifacial microsomia and Treacher-Collins Syndrome [18,19]. MDO provides a less invasive alternative to procedures with higher