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COMMENTARY

Understanding the Spectrum of Treatment Options for Infants With Pierre Robin Sequence and Airway Obstruction

Commentary on Ehsan et al. Longitudinal sleep outcomes in neonates with Pierre Robin sequence treated conservatively. *J Clin Sleep Med.* 2019;15(3):477–482.

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Pierre Robin sequence (PRS), also referred to as Robin sequence, is defined by micrognathia, glossoptosis, and upper airway obstruction and results in varying degrees of airway compromise after birth.[1,2] Both evaluation and treatment of airway obstruction in infants with PRS varies between centers with considerable controversy around the role of polysomnography for the diagnosis of sleep-related breathing disorders and assessment of treatment response.[3,4] Disagreements about the role of polysomnography are undoubtedly influenced by access to polysomnographic testing as this is limited or unavailable in many areas.[5,6] On the treatment side, there are differences between centers with respect to available treatment options and how treatment success is evaluated. While there has been considerably more study of the surgical options compared to nonsurgical options, the field overall is hampered by a relative paucity of high quality evidence.[7]

There is a relatively long list of options for management of airway obstruction in infants with PRS. Nonsurgical options include prone sleep position,[8] supplemental oxygen, use of an oral appliance with a velar extension,[9] oropharyngeal or nasopharyngeal tube, noninvasive ventilation (typically continuous positive airway pressure), and placement of an endotracheal tube.[3] Surgical options include tongue-lip adhesion (or glossopexy), mandibular distraction osteogenesis, subperiosteal release of the floor of the mouth, and tracheostomy—with additional surgical options used more selectively.[3] There are multiple algorithms from different centers to support treatment decisions but none incorporate all treatment options and all use different strategies for decision making.[3,10,11] With few studies comparing treatment modalities and even fewer randomized trials, there is little information on the characteristics of infants that will best respond to any given therapy.[7,11] The study by Ehsan and colleagues in this issue of the *Journal of Clinical Sleep Medicine*[12] adds another option, watchful waiting, to this list.

The study reports on a cohort on infants with PRS who were treated with conservative (ie, nonsurgical) management.[12] The study design was a 14-year retrospective cohort study that included infants with PRS who underwent polysomnography under 3 months of age, had a repeat polysomnography before 12 months of age, and who were managed conservatively at a single center. The study highlights important challenges to advancing our understanding of treatment options for airway obstruction in infants with PRS. The study was conducted in a center where a multi-disciplinary team that included pediatric pulmonology and sleep medicine experts assessed all infants with PRS, and all infants underwent overnight polysomnography, bedside nasopharyngoscopy, and computed tomography of the maxillofacial skeleton. From their retrospective, 14-year review, the authors excluded 33% of infants from the study group because polysomnography was not completed in infancy. The majority of infants with PRS were managed surgically with the minority (36%) receiving conservative management; this reflects a growing trend for surgical management despite reports that prone sleep position resolves airway obstruction in 40% to 70% of infants with PRS.[3] Conservative management for this cohort included supplemental oxygen and watchful waiting. Follow-up showed that by 1 month of age, the majority of infants achieved full oral feeds and both obstructive and total respiratory events decreased on follow-up polysomnography. They observed changes in sleep parameters that reflected expected changes in healthy infants. This shows that even in the absence of surgical intervention to augment the airway, both feeding and airway obstruction improve. Some of the improvements related to surgical interventions may be attributable to normal developmental improvements in sleep and breathing during the time between a baseline polysomnography and a post-surgical follow-up study.

The study highlights the importance of considering multiple outcomes, including feeding and growth, in addition to the results of polysomnography. Improvement in growth, leading to growth of the airway, is likely an important component of improving airway obstruction in infants with PRS and explains why more infants than older children will see improvement in their obstructive sleep apnea to the point that treatment can be stopped.[13] Airway obstruction compromises breathing and oral feeding, so progression to oral feeding is another marker of improvement in airway obstruction. Feeding difficulties result

in longer length of stay, so early assessment of feeding is important.[14] Assessment of feeding, along with polysomnography, is an important part of a complete initial airway assessment of infants with PRS.[3,15,16]

How to choose the best intervention for an individual infant with PRS is an unanswered question. While randomized controlled trials would provide the highest level of evidence, their application in this setting is challenging given, for example, differences in the expertise with each interventions between centers that might impact treatment outcomes. Multicenter registries of infants with PRS would provide important information if initial and follow-up assessments could be standardized and include objective measures where possible. Standardizing assessment tools, again with a focus on objective measures where possible, as well as including comparison groups would strengthen the quality of evidence derived from single center observational studies and facilitate combining data across studies. Understanding the outcomes that are important to children with PRS and their parents and caregivers will ensure that study results are relevant to clinical care. With considerable heterogeneity between infants with PRS, information from a large number of infants will be necessary to identify consistent characteristics that predict treatment response and favorable health outcomes.

CITATION

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DISCLOSURE STATEMENT

Dr. MacLean reports no conflicts of interest.