

READER'S FORUM

Yang IH, Chung JH, Lee HJ, Cho IS, Choi JY, Lee JH, Kim S, Baek SH

Characterization of phenotypes and predominant skeletodental patterns in pre-adolescent patients with Pierre–Robin sequence.

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Pierre–Robin sequence (PRS) is a rare disease condition, which requires long-term interdisciplinary care from birth, and the role of orthodontists is critical. This is the first study to investigate the dental phenotypes, oral manifestations, skeletodental pattern of the maxillo-mandibular complex, and associated anomalies in Korean pre-adolescent patients with PRS. I appreciate the authors for their dedication and hard work. Although the number of samples is limited and it is an area that requires a lot of further research, I would like to ask your opinion based on the authors' expertise and skilled experience in multidisciplinary collaborative care for patients with PRS.

Q1. The neonates with PRS need immediate care such as mechanical ventilation assistance (continuous positive airway pressure), surgery (tongue lip adhesion, or mandibular distraction osteogenesis), or orthodontic airway plate. I wonder what kind of early intervention the sample in this study received, and whether there would be any difference in skeletal patterns depending on the type of early intervention.

Q2. In the literature, PRS patients are known to have

catch-up growth. Although there is no consensus as to when catch-up growth prevails, some reported that this type of growth could not be seen after 5 years of age. This study reported approximately 35% of the sample had normal mandibular body length ratio or Class I maxillo-mandibular relationship. Could this results be interpreted in terms of catch up growth?

Q3. Although soft tissue evaluation was not included in this study, an interesting aspect of the lateral cephalometric analysis of patients with PRS would be the airway and tongue. Has the posteriorly positioned tongue (glossoptosis) been generally altered or maintained during the pre-adolescent stage? Could you give us a brief comment on the tongue position of preadolescent PRS patients?

Questioned by

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A1. Treatment modalities for PRS patients differs according to severity of airway obstruction (i.e., O₂-saturation, parameters of polysomnography), feeding problems, and age/growth status of patients. In several previous case reports and articles, there have been diverse treatment modalities including positioning, nasopharyngeal airway, mechanical ventilation assistance (continuous positive airway pressure), orthodontic airway plate, and surgical intervention (i.e., tongue lip adhesion, mandibular distraction osteogenesis, tracheostomy) can be used.¹

In neonates with PRS, since use of orthodontic airway plate might have a possibility of reducing the

oropharyngeal airway, I do not recommend to use it. In addition, due to surgical morbidity, the mandibular distraction osteogenesis for neonates with PRS is spared for cases of the intractable airway maintenance.

Because large portions of PRS patients investigated in this study were not born in Seoul National University Hospital, we do not have enough medical records about the kinds of early intervention for airway establishment. Therefore, it is hard to tell whether there would be any difference in skeletal patterns depending on the type of early intervention. We would like to investigate this issue in a form of multi-center-based long-term follow-up study with increased sample size and hope to have a chance to report the results in the future.

A2. Since we do not take lateral cephalograms in new born babies with PRS, it is difficult to investigate whether there would be catch-up growth of the mandible between new-born initial status and around 5 years of age. However, some portion of PRS babies might present catch-up growth of the mandible. In a survey study for European clinicians, van Lieshout et al.² reported that three-quarters of the respondents noted the presence of catch-up growth in their patient population.

In a systematic review of catch-up growth in PRS patients, Purnell et al.³ reported that a minority of objective studies suggested increase in the mandibular growth rate in isolated PRS and that only even fewer studies suggested that the maxillomandibular discrepancy in PRS completely resolves. This kind of research needs a sophisticated study design with longitudinal follow-up, large sample size, and sex- and age-matched control group to draw a statistical difference.

A3. Because the airway problem and tongue position are closely related with each other, it is one of the important topics in clinical study on growth of PRS patients. According to Yang et al.,⁴ predominant skeletodental patterns in preadolescent PRS patients included Class II relationship (57.7%), posteriorly positioned maxilla (76.9%) and mandible (92.3%), hyper-divergent pattern (92.3%), high gonial angle (65.4%), small mandibular body length to anterior cranial base ratio (65.4%), linguoversion of the maxillary incisors (76.9%), and linguoversion of the mandibular incisors (80.8%). These findings suggest that altered tongue position (glossoptosis) in preadolescent PRS patients might be maintained compared to normal

orthodontic patients. We would like to investigate this issue using a longitudinal follow-up study and hope to have a chance to report the results in the future.

If catch-up growth of mandible occurs and the size of the mandible increases near to normal values, tongue might reposition into normal position in preadolescent PRS patients. However, since there are numerous factors which can influence the tongue position, clinicians should be careful to interpret the results from two-dimensional radiograph study and consider the habits and swallowing patterns.

Replied by

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